MENTAL RETARDATION
ABSTRACTS

VOL. 9, No. 3

JULY—SEPTEMBER 1972

U. S. Department of Health, Education, and Welfare
Social and Rehabilitation Service
Rehabilitation Services Administration
Division of Developmental Disabilities
Washington, D.C. 20201

Mental Retardation Abstracts is a quarterly publication of the Division of Developmental Disabilities, Rehabilitation Services Administration. It is a specialized service designed specifically to meet the needs of investigators and other workers in the field of mental retardation for rapid and comprehensive information about new developments and research results and to foster maximum utilization of these results.

Mental Retardation Abstracts is prepared under contract with Herner Information Services, Inc.

Communication concerning the abstracts (except orders and mailing changes to the Superintendent of Documents) should be addressed to:

Ms. Mary Herner
Project Director
Herner Information Services, Inc.
2100 M Street, N.W.
Washington, D. C. 20037

Ms. Patricia Thoben
Project Officer
Division of Developmental Disabilities
Rehabilitation Services Administration, SRS
Washington, D.C. 20201

Reprints are available by writing directly to the author (s) of the article.

Copies of Annotated Bibliographies are not reprinted for distribution.

Use of funds for printing this publication approved by the Director of the Bureau of the Budget, June 30, 1969.

For sale by the Superintendent of Documents, U.S. Government Printing Office, Washington, D.C. 20402
Price $2.25 (single copy). Subscription Price: $8 per year; $2 additional for foreign mailing.

MENTAL RETARDATION ABSTRACTS

Volume 9, Number 3 July–September 1972

TABLE OF CONTENTS

Hearing Impairment and the Mentally Retarded:
A Bibliography of Recent Publications 1
ABSTRACTS 7
BROAD ASPECTS OF MENTAL RETARDATION 7
MEDICAL ASPECTS 14
Diagnosis (General) 14
Prevention and Etiology (General) 28
Etiologic Groupings 37
Infections, intoxication, and hemolytic disorders 37
Trauma or physical agents 65
Disease or disorders of metabolism, growth, or nutrition 70
New growths 109
Prenatal influence 109
Gross brain disease (postnatal) 126
Psycho-environmental 132
Convulsive disorders 135
Chromosomal 140
Miscellany 161
DEVELOPMENTAL ASPECTS 164
Physical 164
Mental 165
Social and Emotional 175
Psychodiagnostics 179
TREATMENT AND TRAINING ASPECTS 186
Educational 186
Psycho-social 198
Occupational 204
Therapy 210
PROGRAMMATIC ASPECTS 217
Planning and Legislative 217
Community 222
Residential 229
Recreational 238
FAMILY 240
PERSONNEL 246
AUTHOR INDEX 253
SUBJECT INDEX 262

ACKNOWLEDGEMENTS

The publishers of the following journals have
granted permission to reprint abstracts and summaries:

Acta Neuropathologica
Acta Paedopsychiatrica
American Journal of Diseases of Children
American Journal of Psychiatry
American Journal of Sociology
American Sociological Review
Biologia Neonatorum
British Journal of Psychology
British Journal of Social & Clinical Psychology
British Medical Journal
California Mental Health Research Digest
Canadian Medical Association Journal
Child Development
Developmental Medicine & Child Neurology
Dissertation Abstracts
Education and Training of Mentally Retarded
German Medical Monthly
Helvetica Paediatrica Acta
International Journal of Neuropsychiatry
Journal of Applied Behavior Analysis
Journal of Clinical Endocrinology and Metabolism
Journal of Comparative and Physiological Psychology
Journal of Consulting and Clinical Psychology
Journal of Educational Psychology
Journal of Experimental Psychology
Journal of Medical Genetics
Journal of the American Medical Association
Journal of the Experimental Analysis of Behavior
Lancet
New England Journal of Medicine
Nursing Research
Panminerva Medica
Perceptual and Motor Skills
Psychological Bulletin
Psychological Monographs
Psychological Record
Psychological Reports
Psychological Review
Rehabilitation Counseling Bulletin
Rehabilitation Literature
Science
Training School Bulletin

HEARING IMPAIRMENT AND THE MENTALLY RETARDED: A BIBLIOGRAPHY OF RECENT PUBLICATIONS

compiled by Alfred Hirshoren, Ed.D., and Lyle L. Lloyd, Ph.D.

In 1971 the Joint Committee on the Deaf Retarded – a joint committee of the American Association on Mental Deficiency (AAMD) and the Conference of Executives of American Schools for the Deaf (CEASD) – perceived the need to assemble a bibliography of published materials concerned with the dual handicaps of deafness and mental retardation which would be useful to the members of AAMD and CEASD, as well as to other organizations and individuals concerned with these paired problems. In response to this need, the compilers have assembled an extensive listing of over 600 publications which are relevant both to hearing impairment and to mental retardation. The listing includes materials published from the early 1900's onward. From this extensive listing, the compilers have selected approximately 100 citations covering a two and one-half year period (materials published in 1970 and thereafter) for the following bibliography of recent publications in this subject area.

It is anticipated that the total bibliography – too lengthy to be presented in its entirety in *Mental Retardation Abstracts* – will be published separately by the AAMD at some time in the near future. When the bibliography is issued, its availability will be announced in *Mental Retardation Abstracts* as well as in other appropriate journals.

ACCREDITATION COUNCIL FOR FACILITIES FOR THE MENTALLY RETARDED. *Standards for Residential Facilities for the Mentally Retarded.* Chicago, Joint Commission on Accreditation of Hospitals, 1971.

ADLER, E. P., ed. *Research Trends in Deafness, State of the Art.* Washington, D. C., Department of Health, Education, and Welfare, 1970.

AMES, M. D.; PLOTKIN, S. A.; & WINCHESTER, R. A. Central auditory imperceptions: A significant factor in congenital rubella deafness. *Journal of the American Medical Association,* 213:419-421, 1970.

ANDERSON, H.; LINDSTEN, J.; & WEDENBERG, E. Hearing defects in males with sex chromosome anomalies. *Acta Oto-Laryngologica,* 72:55-58, 1971.

ANDERSON, R. M.; & STEVENS, G. D. Policies and procedures for admission of mentally retarded deaf children to residential schools for the deaf. *American Annals of the Deaf,* 115:30-36, 1970.

BARNET, A. G. EEG audiometry in children under three years of age. *Acta Oto-Laryngologica,* 72:1-13, 1971.

BERGER, S. A clinical program for developing multimodal language responses with atypical deaf children. In: McLean, J. L.; Yoder, D. E.; and Schiefelbusch, R. L. (eds.) *Developing Strategies for Language Intervention with the Retarded.* Baltimore, Md., University Park Press, 1972, in press (also published as Parsons Demonstration Project Report No. 95).

BERGER, S. Systematic development of communication modes. *Report of the Proceedings of the 45th Meeting of the Convention of American Instructors of the Deaf.* Little Rock, Arkansas, Arkansas School for the Deaf, 1971.

BLUE, C. M. Influence of mode of presentation, age and intelligence on paired-associates learning. *American Journal of Mental Deficiency,* 74:527-532, 1970.

BOHLMANN, H. G.; & HAVERS, W. Exomphalos-makroglossie-gigantismus-syndrom (Weidemann–

Beckwith-Syndrom) bei drei geschwistern (Exomphalos macroglossia gigantism syndrome (Wiedemann-Beckwith Syndrome) in three sisters). *Archiv fur klinische und experimentelle ohren-, nasen und Kehlkopfheil-kunde*, 183(2): 175-182, 1971.

BUNCH, G. O. An academic-vocational program for multiply handicapped deaf students. *Volta Review*, 73(7):417-425, 1971.

BURROWS, N. L.; & LLOYD, L. L. Programming considerations for the deaf-retarded. *Report of the Proceedings of the 45th Meeting of the Convention of American Instructors for the Deaf.* Little Rock, Arkansas, Arkansas School for the Deaf, 1971.

CORTESI, M.; & PATRIARCA, P. L. Cromosoma 18 ad anello. Descrizione di un Caso e Massengna della letterature (Ring-type chromosome 18. Description of a case and review of the literature). *Minerva Pediatrica*, 23(20):902-908, 1971.

CRAMMATTE, A. B., ed. Dept. of Health, Education, & Welfare. *Multiply Disabled Deaf Persons: A Manual for Rehabilitation Counselors.* Washington, D. C., U. S. Government Printing Office, 0-381-018, 1970.

DAHLE, A.; & DALY, D. Influence of verbal feedback on auditory discrimination test performance of mentally retarded children. *American Journal of Mental Deficiency*, 76:586-590, 1972.

DARNELL, W. T. *Comprehensive Programming for the Deaf-Retarded Within New York State: A Survey and Proposal.* Albany, New York, New York State Temporary Commission to Study Problems of the Deaf, 1971.

EWERTSEN, H. W.; & NIELSEN, S. S. Food audiometry. Preliminary report. *Acta Oto-Laryngologica*, 69:457-458, 1970.

FENZ, W. O.; & McCABE, M. W. Habituation of the GSR to tones in retarded children and non-retarded subjects. *American Journal of Mental Deficiency*, 75:470-473, 1971.

FOREHAND, R.; & BAUMEISTER, A. A. Effects of variations in auditory-visual stimulation on activity levels of severe mental retardates. *American Journal of Mental Deficiency*, 74:470-474, 1970.

FORREST, J. M.; & MENSER, M. A. Congenital rubella in school children and adolescents. *Archives of Disease in Children*, 45:63-69, 1970.

FORREST, J. M.; MENSER, M. A.; & BURGESS, J. A. High frequency of diabetes mellitus in young adults with congenital rubella. *Lancet*, 2(7720):332-334, 1971.

FRANCO, S. A.; RILEY, H. D., JR.; & CHITWOOD, L. A. The congenital rubella syndrome. *Southern Medical Journal*, 63:825-830, 1970.

FULTON, R. T.; & LAMB, L. E. Acoustic impedance and tympanometry with the retarded. A normative study. *Parsons Research Center Reports No. 1.* Parsons, Kansas, State Hospital and Training Center, 1970.

FULTON, R.; & SPRADLIN, J. Effects of practice on independent variables on the SISI scores with normal subjects. *Journal of Speech and Hearing Research*, 1971 (also published as Parsons Demonstration Project Report, No. 93).

HALL, S.; & CONN, T. Current trends in services for the deaf retarded in schools for the deaf and residential facilities for the mentally retarded. *Report of the Proceedings of the 45th Meeting of the Convention of American Instructors of the Deaf.* Little Rock, Arkansas, Arkansas School for the Deaf, 1971.

HALL, S.; & TALKINGTON, L. Evaluation of a manual approach to programming for deaf retarded. *American Journal of Mental Deficiency*, 75:378-380, 1970.

HALL, S. M.; & TALKINGTON, L. W. Learning by doing: A unit approach for deaf retarded. *Mental Retardation Research Series No. 18.* Austin, Texas State School, 1970.

HALL, S.; & TALKINGTON, L. The use of resident aides in a speech and hearing program for mentally retarded. *Mental Retardation*, 8:37-38, 1970.

HAMBURG, A. Norrie's disease. *Ophthalmologica*, 160:375-377, 1970.

HAVILAND, R. T. A stimulus to language development. *Mental Retardation*, 10:1-21, 1972.

HEGRENES, J. R.; MARSHALL, N. R.; & ARMAS, J. A. Treatment as an extension of diagnostic function: A case study. *Journal of Speech and Hearing Disorders*, 35:182-187, 1970.

HEUSDEN, A.; WILLEMS, C.; LAMBOTTE, C.; HAINAUF, H.; CHAPELLE, P.; & MALCHAIR, R. Methemoglobinemie hereditaire avec arrieration mentale. Etude de trois nouveaux cas (Hereditary methemoglobinemia with mental retardation. Study of three new cases). *Archives Francaises de Pediatrie*, 28(6):631-645, 1971.

HINE, W. D. Verbal ability and partial hearing loss. *Teacher of the Deaf*, 68:450-459, 1970.

HOGAN, D. D. Cortical responses of retardates for AER audiometry. *American Journal of Mental Deficiency*, 75:474-477, 1971.

HOLM, V.; & THOMPSON, G. Selective learning loss: Clues to early identification. *Pediatrics*, 47:447-451, 1971.

HOMES, L. B. Norrie's disease—an X-linked syndrome of retinal malformation, mental retardation and deafness. *New England Journal of Medicine*, 284:367-368, 1971.

HSIA, Y. E.; BRAUTU, M.; & HERBORDT, A. Genetics of the Meckel syndrome (Dysencephalia Splanchnocystica). *Pediatrics*, 48(2):237-247, 1971.

HURDER, W. P. *Overview of Research and Education of Handicapped Children: The U. S. A. in the Sixties.* Urbana, Illinois, IREC, 1970.

KELLAS, G.; & BAUMEISTER, A. A. Effect of adaptation level on response speed of normal and retarded individuals. *American Journal of Mental Deficiency*, 74:533-536, 1970.

KOPITS, I. Observations on 750 geropsychiatric patients. *Journal of the American Geriatric Society*, 18:353-364, 1970.

LAMB, L. E.; & NORRIS, T. W. Relative acoustic impedance measurements with mentally retarded children. *American Journal of Mental Deficiency*, 75:51-56, 1970.

LAMBOTTE, C.; CARLIER, G.; FREDERIC, J.; & KEUTGEN, J. Presentation d'un cas de translocation C-G caryotype 46,XY,T (Cq- – Gq+) – Parents 46,XX et 46,XY (Presentation of a case of C-G translocation karyotype 46,XY,T (Cq- – Gq+) – Parents 46,XX and 46,XY). *Acta Paediatrica Belgica*, 25(2):119-126, 1971.

LEHRKE, R.; THELEN, T.; & LEHRKE, R., JR. Syndrome associated with group-D chromosome deletions. *Lancet*, 2(7715):98-99, 1971.

LLOYD, L. L. AAMD-CEASD joint committee on the deaf retarded: A committee in action seeks help from the membership. *Mental Retardation*, 9:41, 1971.

LLOYD, L. L. Audiologic aspects of mental retardation. In: Ellis, N. ed., *International Review of Mental Retardation.* New York, Academic Press, Inc., 4:311-374, 1970.

LLOYD, L. L. Behavioral audiometry. In: Ventry, L.; Chaiklin, J.; and Dixon, R., eds. *Hearing Measurement: A Book of Readings.* New York, Appleton-Century-Crofts, 1971.

LLOYD, L. L. Bibliographic material on the dual handicaps of hearing impairment and mental retardation: A progress report of your AAMD-CEASD joint committee on the deaf retarded. *Mental Retardation*, 10:18, 1972.

LLOYD, L. L. The establishment of standards for speech pathology and audiology in facilities for the retarded. *ASHA, Journal of the American Speech and Hearing Association*, 13:607-610, 1971.

LLOYD, L. L. Evaluation programming for the hearing-impaired mentally retarded. In: Flanagan, P. J.; Heber, R. F.; and Rybak, W. S., eds. *Programming for the Multiple Handicapped Mentally Retarded.* Madison, Wisconsin: Rehabilitation Research and Training Center in Mental Retardation, Research Monograph No. 4, 1971.

LLOYD, L. L. You've come a long way baby, but—. *Mental Retardation*, 10:2, 1972.

LLOYD, L. L.; & COX, B. P. Programming for the audiologic aspects of mental retardation. *Mental Retardation*, 10:22-26, 1972.

LLOYD, L. L.; & CROSBY, K. G. Establishment of standards for speech pathology and audiology services in facilities for the retarded. *Mental Retardation*, 10:30-31, 1972.

LLOYD, L. L.; & FULTON, R. T. Audiology's contribution to communication programming with the retarded. In: McLean, J. L.; Yoder, D. E.; and Schiefelbusch, R. L., eds. *Language Intervention with the Retarded: Developing Strategies.* Baltimore, Maryland, University Park Press, 1972, in press.

LLOYD, L. L.; & MOORE, E. J. Audiology. In: Wortis, J., ed. *Mental Retardation: An Annual Review (Vol. 4).* New York, Grune and Stratton, 1972.

LOBB, H. Frequency vs. magnitude of GSR in comparisons of retarded and nonretarded groups. *American Journal of Mental Deficiency*, 75(3):336-340, 1970.

LOCKAMYIER, J. S. On developing the auditory discrimination skills of mentally retarded children. *Journal of Special Education of the Mentally Retarded*, 7:196-197, 1971.

MANIGLIA, A. J.; WOLFF, D.; & HERQUES, A. J. Congenital deafness in 13-15 trisomy syndrome. *Archives of Otolaryngology*, 92:181-188, 1970.

MARTINS, G. The present program at Milton. *Report of the Proceedings of the 45th Meeting of the Convention of American Instructors of the Deaf.* Little Rock, Arkansas, Arkansas School for the Deaf, 1971.

MATTHEWS, J. Communication disorders in the mentally retarded. In: Travis, L. E., ed. *Handbook of Speech Pathology and Audiology* (Revised ed.). New York, Appleton-Century-Crofts, 1971, pp. 801-818.

MILLER, M. H.; & RABINOWITZ, M. Conditioned oriented reflex audiometry with maternal rubella children. *Journal of Communicable Diseases*, 3:59-64, 1970.

MITRA, S. Educational provisions for mentally retarded deaf students in residential institutions for the retarded. *Volta Review*, 72:225, 1970.

MITRINOWICZ-MODRZEJEWSKA. Selected problems of deafness in childhood. *Pediatrica Polska*, 45:9-19, 1970. (In Polish).

MITRINOWICZ-MODRZEJEWSKA. Familial auditory disability of children. *Folia Phoniatrica* (Basel), 22:58-62, 1970.

MOORE, M. V. Speech, hearing, and language in De Lange syndrome. *Journal of Speech and Hearing Disorders*, 35:66-69, 1970.

MOSKOWITZ, H.; & LOHMANN, W. Auditory threshold for evoking an orienting reflex in mongoloid patients. *Perceptual and Motor Skills*, 31:879-882, 1970.

MOTTA, G.; FACCHINI, G. M.; & D'AURIA, E. Objective conditioned-reflex audiometry in children. *Acta Oto-Laryngologica Supplement, 273,* 1970.

OSBORNE, K. M.; BELLEFLEUR, P. A.; & BEVAN, R. C. An experimental diagnostic teaching clinic for multiply handicapped deaf children. *Exceptional Children,* 37:387-388, 1971.

OWENS, M.; & HARPER, B. *Sign Language: A Teaching Manual for Cottage Parents of Non-Verbal Retardates.* Pineville, La., Pinecrest State School, 1970.

PICKERING, D.; LASKI, B.; MACMILLAN, D.; & ROSE, V. 'Little Leopard' syndrome. *Archives of Disease in Children,* 46:85-90, 1971.

PORTER, S. The exceptional unit at the Arkansas school for the deaf. *Report of the Proceedings of the 45th Meeting of the Convention of American Instructors of the Deaf.* Little Rock, Arkansas, Arkansas School for the Deaf, 1971.

POWER, D.; & QUIGLEY, S. *Problems and Programs in the Education of Multiply Disabled Deaf Children.* Urbana Institute for Research on Exceptional Children, University of Illinois, 1971.

PREDESCU, V.; CHRISTODORESCU, D.; TAUTU, C.; & CONSTANTINESCU, E. (Down syndrome with normal karyotype in the blood) Neurologia, Psihiatria, Neurochirurgia, *Romanian Medical Review,* 15(1):41-43, 1970. (In English Jan-Mar 1971).

PRESLAND, J. Who should go to E.S.N. schools. *Special Education,* 59:11-16, 1970.

RAPIN, I.; RUBEN, R. J.; & LYTTLE, M. Diagnosis of hearing loss in infants using auditory evoked responses. *Laryngoscope,* 80:712-722, *1970.*

RICHARDSON, D. L.; SEELY, J. R.; THOMPSON, W. M.; & CHITWOOD, L. A. Combined trisomy-18 and congenital rubella. *Southern Medical Journal,* 64(8):923-924, 1971.

RITTMANIC, P. A. The mentally retarded and mentally ill. In: Rose, D., ed. *Audiological Assessments.* Englewood Cliffs, N. J., Prentice-Hall, 1971, p. 369-404.

ROSS, M.; & LERMAN, J. A picture identification test for hearing-impaired children. *Journal of Speech and Hearing Research,* 13:44-53, 1970.

SALLADE, J. B.; SIEGENTHALER, B. M.; & TARDINBUNO, J. S. Speech-hearing measurements in a mentally normal to below normal group of children. *American Journal of Mental Deficiency,* 1972. (In press)

SANZONE, J. Programmed lesson series in auditory discrimination. *Education and Training of the Mentally Retarded,* 5:117-185, 1970.

SHIPLEY, T. Evoked brain potentials and sensory interaction in the retarded child. *American Journal of Mental Deficiency,* 74:517-523, 1970.

SIEGENTHALER, B. M.; SALLADE, J. B.; & TARDIBUONO, J. S. Speech-hearing measurements in an intellectually average to below average group of children. *American Journal of Mental Deficiency,* 76:427-433, 1972.

STRAMANIS, J., JR.; SHAGASS, C.; & OVERTON, D. Evoked responses in Down's syndrome of young adults. *Electroencephalography and Clinical Neurophysiology,* 29:324-327, 1970.

STREISSGUTH, A. P.; VANDERVEER, B. B.; & SHEPARD, T. H. Mental development of children with congenital rubella syndrome: A preliminary report. *American Journal of Obstetrics and Gynecology,* 108:391-399, 1970.

TAGUCHI, K.; GOODMAN, W.; & BRUMMETT, W. Evoked response audiometry in mentally retarded children. *Acta Oto-Laryngologica,* 70:190-196, 1970.

TALKINGTON, L. Use of the MRP in depicting program needs. *Mental Retardation Research Series, No. 11.* Austin, Texas State School, 1970.

VINCE, D. J. The hospital incidence and clinical significance of congenital heart malformations resulting from rubella embryopathy. *Canadian Medical Association Journal*, 102:374-376, 1970.

WEAVER, R. M. Audiological assessment of the mentally retarded. *Research Relating to Children*, 26:34, 1970.

WERTELECKI, W.; & GERALD, P. Clinical and chromosomal studies of the 18q-syndrome. *Journal of Pediatrics*, 78:44-52, 1971.

YANAGISAWA, S.; & HIRAOKA, K. Familial C-G translocation in three relatives associated with severe mental retardation, short stature, unusual dermatoglyphics and other malformations. *Journal of Mental Deficiency Research*, 15(2):136-146, 1971.

(ZACCHIAS, P.); CRANEFIELD, R. F.; & FEDERN, W. Paulus Zacchias on mental deficiency and on deafness. *Bulletin of the New York Academy of Medicine*, 46:3-21, 1970.

BROAD ASPECTS OF MENTAL RETARDATION

1541 **CZEIZEL, A.; & REVESZ, C.** Major malformations of the central nervous system in Hungary. *British Journal of Preventive and Social Medicine*, 24(4):205-222, 1970.

An epidemiologic survey of 1,315 cases and a family study of 534 cases reveals the incidence of major central nervous system malformations in Budapest (Hungary) from 1963-1967 to be 3.71 per 1,000 births, a rate which exceeds the international rate. Malformations were divided into 4 categories: anencephaly, spina bifida, encephalomeningocele, and congenital hydrocephalus. The rate for tube defects was 2.95 per 1,000 births. There was a marked bias in female anencephaly (67.5%). The incidence of anencephaly and spina bifida was significantly higher in first-born, and the incidence in sibs was 10.3 times the population incidence. (46 refs.) - *G. S. Bell.*

National Institute of Public Health
Budapest, Hungary

1542 **KUSHNICK, THEODORE.** Medical aspects of mental retardation in the year 2000 – which road to take. *Welfare Reporter*, 21(4):3-7, 1970.

If a mature population became involved spiritually, physically, and financially with MR, many exciting events might become a reality by the year 2000. Appropriate treatments for newborns, adolescents, and pregnant women with biochemical genetic defects might be available; immunological techniques might be available for translocation chromosome carriers who are normal physically and mentally but who have a high risk for abnormal offspring; the utilization of proper nutrition to prevent prenatal and postnatal malnutrition damage to the fetal and infant brain might be standard procedure; and numerous medical and educational therapies might be available to MR individuals. At the opposite pole, the most pessimistic predictions about MR in the year 2000 include the continuing explosion of the population, nationwide financial difficulties, and the loss of human qualities necessary for survival. The most realistic prediction posits an increasing but still small number of dedicated individuals shaping the optimum course and expanding the knowledge and care of MR persons. (No refs.) - *B. J. Grylack.*

New Jersey College of Medicine & Dentistry
Newark, New Jersey

1543 **FEINGOLD, MURRAY; FAIN, TOBY; & GELLIS, SYDNEY S.** State-wide information center for handicapped children. *Journal of Pediatrics*, 77(5):830-832, 1970.

A total of 697 inquiries to a pilot program information center for facilities for handicapped children in Massachusetts were followed up in terms of source of the inquiry and utility of the information. Over half the callers (52%) were parents. Friends and agencies also used the service, but only 4% of the callers were physicians. Most callers sought information about home programs for the handicapped. Eighty-four percent of callers acted upon the information, and 80% found it useful. Most had been unable to locate the information elsewhere. Because of the success of the 2-year pilot program the Center was incorporated into the State Health Department. A computerized national information center would appear to be the next logical and feasible step. (No refs.) - *E. L. Rowan.*

Boston Floating Hospital for
Infants and Children
Boston, Massachusetts 02111

1544 **KNOX, E. G.; & MAHON, D. F.** Evaluation of 'Infant at Risk' registers. *Archives of Disease in Childhood*, 45(243):634-639, 1970.

An examination of Infant at Risk Registers, utilizing record linkage techniques, has demonstrated the existence of positive associations between assessment of risk and reporting of subsequent defects at age 3 and between assessment of risk based on maternity data and the likely need for special education at age 13. However, the use of these registers for defining predictions appears not to be justified, and they should, instead, serve the purpose of registering known handicapped children and extending surveillance to all children. The results of an analysis of an observation register containing records of identity, clinical observations, and handicap details of all live births in Birmingham over a 2-month period showed that when the observation register was linked with a file on defective children and submitted to the Education Department 3 years later, 3 ambiguities existed: some defects were ascertained, not predicted; other predicted defects were minor and/or had been resolved; and retrospective "predictions" occurred when later observations were entered in the register. Another analysis which linked maternal and social data with a file of the Education Department on children receiving special education showed that while 358 of the 652 records on the Education Handicap file were successfully linked, no record existed of handicaps unlinked for technical reasons, and the problem of migration also made evaluation difficult. Results of both analyses suggest that the bases of prediction were insufficient to warrant special concentration on a particular group and withdrawal of surveillance from another. Observation of every infant's developmental progress is necessary. (4 refs.) - *M. S. Fish.*

The Medical School, Edgbaston
Birmingham 15, England

1545 CHILDERS, RALPH L. Toward a science of rehabilitation. *Journal of Rehabilitation,* 36(6):21-24, 1970.

Rehabilitation, which has done a good job in restoring the physically and mentally handicapped to socially useful roles, must now develop into a science if it is to perform equally successfully with the socially and culturally disadvantaged. Method, style, and generic concepts and theories must be clarified, chiefly by application of the observation, induction, and deduction characteristic of the scientific method. Opponents of such a view are misled by faulty analogies with physics and biology; rehabilitation is now sufficiently advanced to permit formulation of the viable questions that will lead to firm patterns. Determination of the components necessary to a science of rehabilitation will yield a group of 5 basic components: a set of values; a set of hypotheses; a set of methods with which to test the hypotheses; a body of meaningful data; and implementation of findings by implication and use. Rehabilitation is now ready to proceed on this course. (7 refs.) - *B. Berman.*

University of Iowa
Iowa City, Iowa

1546 DORIS, JOHN. Science, action, and values in familial retardation. *Journal of Special Education,* 4(2):161-170, 1970.

Although there is merit in Albee's proposal that the excessive concentration on biomedical research in MR be cut back and a major share of funds be devoted to educational and rehabilitation research, some of the scientific facts and theory on which the proposal is based are of questionable validity. Albee assumes that the familial MR represent the tail end of a distribution determined by the normal interaction of polygenic factors. In the light of present genetic knowledge this assumption is gratuitous because of variation in phenotypic and genotypic distributions. Social action programs based on Albee's model might take the form of selective breeding or radical changes in educational programs. These would be premature and could create more problems than they would solve, as well as discredit and hinder the advance of the behavioral sciences. (12 refs.) - *J. K. Wyatt.*

Cornell University
Ithaca, New York

1547 FREEMAN, ROGER D. The myth of perfectibility. *Journal of Special Education,* 4(2):171-176, 1970.

Albee's thesis that efforts to prevent mild MR are futile, since a certain percentage of the population will always be retarded, is timely and has partial validity. However, he should provide balanced information in his proposal, even

though such data might reduce the force of his argument. Albee presents a simplified version of the complex interaction of polygenic factors with prenatal, perinatal, and postnatal influences. He is frequently unclear as to whether he is referring to phenotype or genotype, and his conclusions regarding the normal distribution of intelligence are premature and misleading. Although Albee seems to assume that biomedical and behavioral approaches have been opposed to each other, the various disciplines should not be dichotomized but should work together as a team to meet individual needs. (20 refs.) - *J. K. Wyatt.*

University of British Columbia
Vancouver, B. C., Canada

1548 **NELSON, THOMAS L.** The case for multidisciplinary research efforts in mental retardation. *Journal of Special Education,* 4(2):177-179, 1970.

Major MR research centers should be truly multidisciplinary; polarization between the biomedical sciences and the behavioral sciences should be avoided. Since a variety of factors contribute to MR, the contributions of a multidisciplinary research team are needed in order to develop relevant treatment and prevention approaches. Although Albee emphasizes that organic pathology is absent in mildly retarded individuals, neuropathological studies show that the majority have organic lesions, indicating the influence of physiological factors. Biomedical research cannot be neglected but must be combined with social, educational, and rehabilitation research. (9 refs.) - *J. K. Wyatt.*

University of California
Irvine, California

1549 **WALTER, DONALD O.** Some logical and mathematical errors in support of a worthy cause. *Journal of Special Education,* 4(2):181-183, 1970.

The reasoning used by Albee to support his conclusions is untrue and filled with logical and mathematical errors. He predicts that because of polygenic factors some 2½% of all children born will be MR and that this prediction will hold true today, tomorrow, next year and for the next century. This prediction is based on the mistaken belief that when a curve is "approximately Gaussian," its shape is unalterable, when in fact a rather small change in shape could make a great change in Albee's figure. Albee oversimplifies when he states that "if the distribution of IQ is approximately a Gaussian curve, then inheritance of the physical basis of intelligent behavior is exclusively polygenic." The approximate Gaussian curve observed for IQ is not strong enough to exclude the value of further important biomedical progress against MR caused by major genes. It is true that more money should be spent on educational research for the MR, but not at the expense of educational research generally. (No refs.) - *J. K. Wyatt.*

University of California
Brain Research Institute
Los Angeles, California

1550 **ALBEE, GEORGE W.** But some are more equal than others! *Journal of Special Education,* 4(2):185-188, 1970.

Although professions should be equal and scientific merit should be the only criterion for federally funded research, in practice, some professions turn out to be more equal than others. Educational research costs money, and the money is being spent for biomedical research. Intelligence does fall into a bell-shaped curve, and there are actually 2 curves. Medical research is urgently needed for the smaller, lower curve, which mainly consists of exogenously damaged persons. Even if every psychiatrist-in-training had experience with MR, the manpower needed to scratch the surface of intervention needs would be lacking. Even if all social and cultural deprivation were eliminated, 2% of all children would still be classified as normally deficient. We do not need exact knowledge of the beneficial effects of intervention programs in order to make decisions about implementing them. Professions differ from sciences in that they exist to serve society's immediate needs and must often make decisions in the absence of specific scientific data. If social and educational opportunities were equalized for all racial and ethnic groups, they would all have the same proportion of MR children. We need collaboration in research, but must it always be in a biomedically-oriented center? (1 ref.) - *J. K. Wyatt.*

Case Western Reserve University
Cleveland, Ohio

1551 **ALBEE, GEORGE W.** Let's change our research priorities in retardation. *Journal of Special Education,* 4(2):139-147, 1970.

Most MR is due to polygenic inheritance factors, and major medical breakthroughs cannot be expected to bring about a significant reduction in incidence. However, at the present time, most research construction funds are used for biomedical research centers aimed at reducing the incidence of the 25% of MR caused by genetic and pathological factors. At least half of available research funds should be used to develop and evaluate services and programs aimed at maximizing limited potential and to provide socially innovative opportunities aimed at the development of reasonable self sufficiency. Most familial MRs do not have an identifiable medical defect. However, psychiatrists, many of whom are disinterested in MR and have minimal training or work experience with MR subjects, treat MR as a medical rather than an educational problem. Treatment should emphasize habilitation or rehabilitation. (16 refs.) - *J. K. Wyatt.*

Case Western Reserve University
Cleveland, Ohio

1552 **BENSON, P. F.,** ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, 224 p.

This compilation of contributions by members of a Study Group held at the Ciba Foundation, London, under auspices of the Institute for Research into MR, focuses on biological phenomena at subcellular and molecular levels approached from widely varying research and technological orientations. The discussion following each presentation illustrates the desirability of a multidisciplinary investigation of biological problems, taking advantage of the fresh insights and mutual gains induced by interchange of ideas. This publication makes the presentations and discussions available to a large audience of biochemists, biologists, clinicians, geneticists, neuropathologists, and pharmacologists working in cognate fields. (530 refs.) - *B. Berman.*

CONTENTS: Membrane Potentials and Metabolic Performance (Bradford); Distribution and Utilisation of Amino Acids in the Brain (McIlwain); Membrane Transport of Amino Acids (Baerlocher, Mohyuddin, & Scriver); Defects in Renal Tubular Reabsorption of Amino Acids (Raine); Hartnup Disease (Jepson); Lysosomes in Nervous Tissue (Dingle); Biochemistry of the Mucopolysaccharidoses (Muir); Aspartylglycosaminuria—A New Inborn Error Possibly Due to a Lysosmal Enzyme Defect (Pollitt & Jenner); Types of Lysosomal Abnormality (Allison); Biochemistry of the Gangliosides in Relation to Inborn Disorders of Ganglioside Metabolism (Myant); Laboratory Diagnosis of Some Sphingolipidoses (Raine); Biochemistry of Down's Syndrome (Stern); RNA Synthesis by Down's Syndrome Leucocytes (Benson); The Effects of Chromosome Redundancy on Gene Expression (Yielding); Chromosome Gain in Higher Plants (Rees & Jones).

1553 **ALDRICH, ROBERT A.; HOLLIDAY, AUDREY R.; COLWELL, DAVID G., JR.; JOHNSON, BETTY JO; SMITH, ELLEN M.; & SHARPLEY, ROBERT P.** The Mental Retardation Service Delivery System Project. A Survey of Mental Retardation Service Usage and Needs among Families with Retarded Children in Selected Areas of Washington State. *Research Report,* 17(3):62 p., 1971.

This survey of 166 parents and guardians of children under 14 years of age who had sought MR services focuses on family composition and demographic information, diagnostic data, information on services requested and/or obtained, general effects of MR on the family's life style, and family attitudes toward MR services. Incidence of MR was higher among the children of women over age 35. MR appeared to be suspected and confirmed at an earlier age in urban-suburban areas than in rural areas, and children in urban-suburban areas had contact with more services. Children from families of higher socioeconomic status were identified earlier and received more services than children from lower socioeconomic status. Families from higher socioeconomic status were more adversely affected by an MR child and spent more for services than those of lower socioeconomic status. Over half the children had moderate to severe speech problems. Of the total group of MR children, 82% were receiving some type of schooling. Cost of MR services was higher for parents of SMRs. (21 refs.) - *J. K. Wyatt.*

University of Washington
Seattle, Washington

1554 **WILLIAMS, PHILLIP; DAVIES, PAT; EVANS, ROY; & FERGUSON, NEIL.** Season of birth and cognitive development. *Nature*, 228(5276):1033-1036, 1970.

It is suggested that changes are needed in the English educational system to reduce the incidence of certain educational and behavioral handicaps among children born during the summer. The summer-born child is 50% more liable than others to have such a handicap, e.g., retarded reading, child guidance case, educationally subnormal. Summer-born children spend less time than others in the infant schools and are usually the youngest in their school age-group; this may be etiologically related to the increased incidence of these handicaps among such children. Some details of the testing of certain children are provided. (22 refs.) - *E. Kravitz.*

The Open University
Bletchley, Buckinghamshire, England

1555 **MUHLETHALER, J. P.** Kongenitale Missbildungen (Congenital malformations). *Helvetica Paediatrica Acta*, Supplement XXIII, 1970. 50 p.

Although in recent years many studies have been made concerning congenital malformations, very few investigations have been made in pathology institutes, where the most comprehensive analysis of mortality rates can be conducted. In the study reported here, a precise investigation of the Argovian (Swiss Kanton) statistical and pathological data on congenital malformations and the subsequent mortality rate for the years 1961-1965 was carried out. Of the malformations which caused death (1.46%), lesions of the cardiovascular system and the nervous system were most common. The remaining cases involving death are attributable to malformations of the alimentary, respiratory, and genito-urinary systems, to congenital hernias and omphaloceles, and to various metabolic defects. A further breakdown of the figures for the most important categories was made. Among these findings it was noted that in comparing the relative number of boys and girls represented in the mortality figures, a preponderance of boys was striking in malformations of the digestive and respiratory tract. Male predominance was also found for diaphragmatic hernia and malformations of the genito-urinary system. No significant difference in the total incidence of malformations among the local population and the immigrated Italian population was observed. (49 refs.) - *P. Custers.*

Pathologisches Institut des Kantonspitals
Aurau, Switzerland

1556 **MERRILL, ROBERT E.** Prognosis for children with multiple handicaps. *American Journal of Diseases of Children*, 121(3):207-212, 1971.

The success rate of multiply handicapped children followed at a clinic was unremarkable and suggests a reevaluation of general support of such patients and their families. The histories of 32 patients whose association with the clinic was terminated by death or by transfer to a clinic for adults constitute the material of the study. Six, or 19%, of these cases may be considered successful, and another 6, or 19%, may have a remote potential for success. Of the remaining 20 patients, or 62%, 9 are dead and 11 are absolute failures. Failure may be due to a lack of medical knowledge in prevention and treatment of disorders, poor environment, or a combination of organic and psychological problems. The results indicate that eventual success depends on good follow-up and on an effective school and vocational plan. Not less necessary are stimulation of motivation at the ideal time and the introduction of imagination and flexibility into school and vocational programs when the child is very young. (2 refs.) - *S. Glinsky.*

University of Virginia School of Medicine
Charlottesville, Virginia 22901

1557 **KHALILI, ABBASS; MARIENFELD, CARL J.; WRIGHT, HARLEY T.; & WEISS, EDWARD S.** An approach to the estimation of the true number of congenital malformations. *Pediatrics*, 46(5):712-720, 1970.

A method for estimating the number of birth defects by matching birth certificate reports describing birth defects with infant death certificates has shown that estimated rates closely

resemble those obtained from the study of hospital records. Computer matching of 6-year records of 4,900 birth certificates showing birth defects and 13,154 punch cards from infant death certificates on which 1,852 were indicated as attributable to congenital malformations showed that the average rate of birth defects in the state under study (Missouri) was 24.5/1000 live births over the 6-year period. Hospital records indicate this rate to be 24.4/1000 live births. By eliminating factors related to reporting, the estimate provides a useful method for determining actual differences between geographic areas and may be especially useful in the study of environmentally caused health effects. (25 refs.) - *M. S. Fish.*

University of Missouri
Environmental Health Surveillance Center
Columbia, Missouri 65201

1558 Voorstel tot de eenmaking van de nomenclatuur en de onderverdeling inzake zzakzinnigheid, gebruikt door de verschillende overehidsinstanties (Proposal to unify the terminology and categorization in relation to imbecility, used by governmental agencies). *Amentia*, 23:16-17, 1971.

The terminology used by different governmental agencies in Belgium for the classification of MR is confused and often not well defined. The proposal designed to remedy this situation is based on the development characteristics of MR children, without specific indications as to potential educability. The suggested new breakdown and terminology closely follows that currently used by the World Health Organization. (No refs.) - *P. Custers.*

1559 Les enfants atteints de handicaps multiples (Children afflicted with multiple handicaps). *Nos Enfants Inadaptes*, 36:8-9, 1970.

The lack of special attention to children with multiple handicaps in France is dramatically illustrated by the complete absence of any useful statistical information as to incidence, and by the almost total inattention to any medical and educational solutions. The present situation is characterized by delayed attention to secondary handicaps in diagnosis of the child, the special burdens placed upon the parents who often have to consult several specialists for help, and the lack of institutional recognition of the special problems involved in the care of these children. From these observations follow recommendations concerning administrative measures, areas of needed medical research, and assistance to parents which could be undertaken to deal adequately with the special problems of children with multiple handicaps. (No refs.) - *P. Custers.*

1560 **PORTAL-FOSTER, C. W.** Community diagnosis: Mental retardation and social science. *Canadian Journal of Public Health*, 62(1):46-53, 1971.

This discussion of epidemiological studies in MR focuses on study population and sample, case finding, and definitional criteria. An ongoing prevalence study of the Prince Edward Island (Canada) populations is cited and a number of guidelines that shaped the study are discussed. (18 refs.) - *B. Berman.*

Canadian Council on Social Development
Ontario, Canada

1561 **SPRINGER, NINFA SATURNINO, ed.** *Proceedings of a Conference on Nutrition and Mental Retardation.* Ann Arbor, Michigan, University of Michigan Publications Distribution Service, 1971, 123 p.

Held on February 10-11, 1971, the Conference on Nutrition and Mental Retardation was sponsored by the Institute for the Study of Mental Retardation and Related Disabilities, one of the University Affiliated Centers established throughout the U.S. and supported by an HEW grant. The conference was planned chiefly to provide nutritionists and dietitians with an overview of MR and the role of nutrition in its prevention and management. The importance of nutrition in this respect is widely recognized for the fetal and early postfetal periods, for the child's normal physical and mental development, and for dealing with MR deriving from inborn errors of metabolism. Feeding technology is an area of great potential significance for health personnel caring for retarded children. (52 refs.) - *B. Berman.*

CONTENTS: (1) Mental Retardation: An Overview; (2) Nutrition Manpower Needs; (3) Nutrition and Learning in Children; (4) Maple Syrup Urine Disease and Phenylketonuria; (5) Feeding Problems of Mentally Retarded Children; (6) Nutritionists in University Affiliated Centers.

1562 **CRUIKSHANK, WILLIAM H.** Mental retardation: An overview. In: Springer, Ninfa Saturnino, ed. *Proceedings of a Conference on Nutrition and Mental Retardation.* Ann Arbor, Michigan, University of Michigan Publications Distribution Service, 1971, p. 3-11.

The problem of MR, which is defined in terms of anything that may negatively affect intellectual function and cause incurable deviation in growth and development, must be approached from a multi- or interdisciplinary orientation. In terms of IQ, MR shows various gradations: profound retardation (IQ 0-25); severe retardation (IQ 25-40); trainable (IQ 40-60); educable (60-80); slow learners (IQ 80-low 90's). All groups pose community and public health responsibilities, both socially and vocationally. MR determinants include genetic and environmental factors, as well as the intellectual apathy that accompanies chronic unemployment. In depressed socioeconomic groups, lack of adequate environmental interplay among children under 3 years of age and nutritional deprivation are serious causes of retardation. There is also the serious impact of neurological and motor problems. Perceptual problems produce visual, audio, tactile, and associated retardation consequences. About 75% of cerebral palsy children fall below IQ 80, and 30% of blind children function at a retarded mental level. (No refs.) - *B. Berman.*

University of Michigan
Ann Arbor, Michigan

1563 **KATZ, ELIAS.** On "Psychology and Mental Retardation" position statement. *American Psychologist,* 26(1):103, 1971.

A multidisciplinary approach is needed to the problems of the MR, who must be seen as a total human being. The collective task should be to assist the MR in achieving his maximum potentials, including personal and social adjustment, physical development, creativity, and educational and vocational competency. (No refs.) - *S. Glinsky.*

Center for Training in Community Psychiatry
and Mental Health Administration
Berkeley, California 94704

MEDICAL ASPECTS – Diagnosis (General)

1564 **AUBRY, RICHARD H.; & NESBITT, ROBERT E. L., JR.** High-risk obstetrics, V. Cytohormonal and interhormonal relationships in normal and abnormal pregnancy. *American Journal of Obstetrics and Gynecology*, 107(7):990-1001, 1970.

Serial estriol (E), pregnanediol (P), chorionic gonadotrophin (HCG), and karyopknotic index (KPI) determinations were made throughout pregnancy in 20 normal controls, 93 high-risk patients with normal outcomes, and 31 high-risk patients with abnormal outcomes. Normal pregnancy is characterized in general by increasing E and P and decreasing HCG and KPI. The most reliable indicator of fetoplacental distress was a decrease in E, and of chronic distress, an increase in the KPI. Despite the reliability of these trends, many false-negative results are noted. (29 refs.) - *E. L. Rowan.*

Upstate Medical Center
Syracuse, New York

1565 **DALTON, NOEL T.** The significance of rhesus antibodies in liquor amnii. *American Journal of Obstetrics and Gynecology*, 107(6):925-932, 1970.

In a series of 111 Rh-isoimmunized pregnancies both the titer of rhesus antibodies and the optical density of amniotic fluid were used to predict fetal outcome. The Rh titer correctly predicted fetal status in 68% of the cases and the optical density in 69%. Rh titer was not affected by maturity or contamination of the liquor by heme pigments or meconium. A titer of 1:64 or greater was associated with fetal death in 12 of 14 cases. It is hypothesized that the common factor in both measurements may be protein binding, but this aspect and the selective transmission of antibodies to the fetus and amniotic fluid require further elucidation. (19 refs.) - *E. L. Rowan.*

Auckland Hospital
Auckland, New Zealand

1566 **ECHT, CHARLES R.; & COHEN, LAWRENCE.** The management of high-risk pregnancies: An evaluation of estriol determinations. *American Journal of Obstetrics and Gynecology*, 107(6):947-953, 1970.

Serial estriol excretion determinations in 122 pregnancies (33 normal, 37 toxemic, 18 prolonged, and 34 diabetic) confirmed earlier reports that an elevated and/or rising level was associated with fetal well-being, while a low and/or falling level meant fetal distress. A urinary estriol level less than 4 mg per 24 hours during the last 6 weeks of gestation almost always was associated with fetal death. Values in the range from 4 to 12 mg per 24 hours, especially when rapidly falling from higher levels, were also highly correlated with fetal wastage. Close monitoring of estriol may be of value in the management of the high-risk pregnancy. (31 refs.) - *E. L. Rowan.*

Indiana University Medical Center
Indianapolis, Indiana

1567 **LEE, TZU Y.; & *WEI, PING Y.** Spectrophotometric analysis of amniotic fluid in anencephalic pregnancies. *American Journal of Obstetrics and Gynecology*, 107(6):917-920, 1970.

Spectrophotometric analysis of the amniotic fluid obtained in 4 pregnancies resulting in the birth of an anencephalic child showed each to have an absorption peak at 450mμ. Two of the mothers had hydramnios and 2 did not. The 450mμ peak had been thought to be specific for Rh-immunized pregnancy; however, it appears to be a bad prognostic sign for the fetus regardless of its cause. (10 refs.) - *E. L. Rowan.*

*1, Chang-Te Street
Taipei, Taiwan, Republic of China

1568 **LEE, C.L.Y.; GREGSON, N. M.; & WALKER, S.** Eliminating red blood-cells

from amniotic-fluid samples. *Lancet*, 2(7667):316-317, 1970. (Letter)

A technique which provides for lysing of erythrocytes and platelets in amniotic-fluid samples is a useful procedure for freeing these samples from contamination without harming other components. The procedure employs ammonium chloride for lysing unwanted cells of the precipitated fraction of centrifuged amniotic fluid. Without removal of these contaminants, the rate of growth of amniotic-fluid cells in cultures is inhibited. Of a series of 24 amniotic-fluid samples obtained by amniocentesis or by hysterotomy at 10-36 weeks' gestational age, 11 were contaminated with blood, and the degree of contamination ranged from 16,000 to 440,000 red blood cells/ml. Comparison of results from the use of the ammonium chloride procedure with those from untreated cultures, both from these latter contaminated samples, showed that the untreated cultures with red blood cells either failed to grow or required several additional days, whereas cultures from treated, contaminated samples showed initial growth as early as did cultures from the uncontaminated samples. This technique should prove useful in expediting diagnostic procedures based on cultured amniotic-fluid cells. (3 refs.) - *M. S. Fish.*

University of Liverpool School of Medicine
Liverpool, England

1569 **COWIE, VALERIE A.** Prenatal diagnosis. *Parent's Voice*, 20(4):15, 1970.

Amniocentesis can be used for prenatal diagnosis of certain genetic conditions associated with MR. The fluid can be examined to detect chromosomal abnormalities and biochemical disorders in the baby. A positive finding permits parents to make decisions regarding termination of pregnancy, while a negative finding provides reassurance to the mother. Although this service is not presently available to the public in the United Kingdom, it is carried out in approximately 6 research centers. (No refs.) - *J. K. Wyatt.*

No address

1570 **MILUNSKY, AUBREY; LITTLEFIELD, JOHN W.; & ATKINS, LEONARD.** Tetraploidy in amniotic-fluid cells. *Lancet*, 2(7680):979, 1970.

The significance of the presence of tetraploidy in cultured amniotic-fluid cells has been under investigation. In a series of 20 consecutive cases for clinical diagnosis, a variable tetraploidy of 0 to 43% was found in cultured amniotic-fluid cells obtained through amniocentesis at gestation of 14 to 16 weeks. Ten of the 20 cases have been delivered and are phenotypically normal infants. It appears at present that the occurrence of tetraploidy, even in most metaphases in amniotic-fluid cell cultures, is consistent with the birth of a child with a normal karyotype. While it is believed that the origin of the cells becoming tetraploid is probably amnion, the explanation for the wide variation in the percentage of tetraploidy is still uncertain. (No refs.) - *B. J. Grylack.*

Massachusetts General Hospital
Boston, Massachusetts 02114

1571 **GALLOWAY, R. K.** Clinical experience with fetal blood pH measurement in fetal distress. *Journal of Obstetrics and Gynaecology of the British Commonwealth*, 77(7):587-590, 1970.

Fetal scalp capillary blood was collected from 81 primigravida and 19 multigravida women during the first stage of labor who showed 1 or more signs of fetal distress at that time. The pH of the blood was measured on an Astrup Radiometer microelectrode in order to facilitate clinical management. In the group of 31 women where the sample to delivery interval was greater than 3 hours, no correlation between pH and Apgar score was found. In the 69 cases where the interval was less than 3 hours, high pH values usually were associated with Apgar scores of 6 or more, whereas a pH of less than 7.21 was associated with a depressed baby. It appears that a pH of 7.25 or greater measured less than 3 hours before delivery usually means a vigorous baby at birth, assuming no change in clinical signs of distress during that interval. No reliance can be placed on the prediction if more than 3 hours elapses between samples or between the last sampling and delivery. The technique is of greater value when combined with continuous fetal heart rate monitoring. (4 refs.) - *B. J. Grylack.*

Royal Infirmary
Edinburgh, Scotland

1572 **HAKIM, C. A.** Placental localization. A comparison of the Doppler ultrasound method with that of radioactive carbon monoxide and the gamma camera. *Journal of Obstetrics and Gynaecology of the British Commonwealth*, 77(7):625-626, 1970.

One hundred obstetrical patients between 28 weeks gestation and term were examined for placental localization by means of the Doptone ultrasonic apparatus and radioactive carbon monoxide using the gamma camera. Of the 50 cases delivered by caesarean section, both methods predicted correctly in regard to the level and position of the placenta in 47. In 49 of the 50 cases delivered vaginally, the 2 methods of localization agreed, but there was no direct confirmation of the placental site. The advantages of the Doptone method as compared with the isotopic method are its simplicity, the ready availability of the instrument in many obstetric units, and the fact that the investigation can be carried out at the bedside of the patient without prior preparation and without the involvement of any other departments. Both techniques are useful in determining the localization of the placenta before amniocentesis and intrauterine transfusion. (4 refs.) - *B. J. Grylack.*

Hammersmith Hospital
London W. 12, England

1573 **DAW, EDWARD.** Intra-uterine fetal visualisation. *Nursing Mirror*, 131(10):14-15, 1970.

Seventy patients with polyhydramnios have been examined for fetal abnormality with the Myodil technique. Several unusual presentations have been observed, but no soft-tissue tumors have been diagnosed. (2 refs.) - *B. J. Grylack.*

Nether Edge Hospital
Sheffield, England

1574 **OKA, EIJI.** Seishin-hakujakuji no noha ni kansuru kenkyu (An electroencephalographic study of mentally retarded children). *Psychiatria et Neurologia Japonica*, 72(6):555-578, 1970.

A new systematization of MR children without clinical epilepsy or cerebral palsy based on abnormalities in EEG patterns, was found to provide one of the strongest indications for the estimation of causes of retardation. The 318 cases were divided into subnormal, pathological, and mixed groups, with the subnormal further subdivided into familial and cryptogenic groups and the pathological into exogenous (limited to Down's syndrome) and endogenous groups. Abnormal EEG patterns were observed in 89% of the cases; slow wave dysrhythmia was found most frequently (in 55.4% of those exhibiting abnormalities) and was presumed to be the most basic EEG abnormality of MR children. Low voltage dysrhythmia, asymmetry, fast wave dysrhythmia, and diffuse hypersynchronous theta patterns can be considered as the characteristic abnormal pattern of the exogenous group. The epileptic discharge was most common in the exogenous group, but was rare in Down's syndrome and the subnormal familial group. In some cases of the subnormal familial group, which could usually be regarded as physiological deviation, abnormal findings such as spikes, asymmetry, and low voltage dysrhythmia were suggestive of some organic brain lesion. This fact suggests the presence in these cases of some unknown, doubly loaded exogenous factor. (47-item bibliog.) - *N. Behling.*

Okayama University
Okayama, Japan

1575 **WALKER, S.; LEE, C.L.Y.; & GREGSON, N. M.** Polyploidy in cells cultured from amniotic fluid. *Lancet*, 2(7683):1137, 1970. (Letter)

Polyploidy was observed in cultured amniotic fluid obtained by amniocentesis or at hysterotomy. Of the 110 cases, 13 showed no polyploidy cells, 61 (55%) had more than 3% polyploids, 25 (23%) had at least 10% polyploids and 3 cases had all polyploidy cells. The chromosome analysis showed primarily tetraploidy. It is suggested that the frequency of polyploidy without concomitant abnormality contraindicates using the presence of polyploidy as an adequate indication for abortion. (1 ref.) - *E. Kravitz.*

University of Liverpool
Liverpool, England

1576 **STEVENS, JANICE R.; & MILSTEIN, VICTOR.** Severe psychiatric disorders of childhood: Electroencephalogram and clinical correlates. *Mental Health Digest*, 2(12):29-33, 1970.

One hundred children admitted during an 8-month period to a children's psychiatric unit of a state hospital were studied to assess the relationship between clinical and EEG features for this type of population. Analysis of the regional distribution of abnormal wave forms recorded from the scalp indicated positive relationships between frontal region abnormality and hyperactivity, parieto-occipital abnormality and history of hallucinations and difficulties with comprehension, and abnormality of the temporal region with history of abdominal pain. The slow EEG seems to relate to disturbances of intellectual and motor development, while the spike-wave at rest and with light is associated with behavioral and emotional instability. The abnormal EEG found in almost half of the children with psychiatric disorders suggests associated biochemical or physiological abnormalities which lead both to characteristic behaviors and to regional disturbances in brain function. (No refs.) - *B. J. Grylack.*

No address

1577 The signs of inability to metabolize vitamins. *Medical World News*, 11(36):4-5, 1970.

Vitamins are not metabolized properly in various childhood diseases, ranging from methyl-malonic aciduria to homocystinuria. Patients with these diseases may present with failure to thrive, protein intolerance, or unexplained acidosis. It is important to make a diagnosis before MR, dislocation of eye lenses, paraplegia, or other serious signs appear. It is possible to screen for these inborn metabolic disorders. At the present time it is known that vitamins B_1, B_2, B_6, and B_{12} cannot be handled by infants with various disorders, and others may be implicated in the future. Therapy requires 100 to 1,000 times the daily normal vitamin need. (No refs.) - *B. J. Grylack.*

1578 **CAMPBELL, STUART.** Ultrasonic fetal cephalometry during the second trimester of pregnancy. *Journal of Obstetrics and Gynaecology of the British Commonwealth*, 77(12):1057-1063, 1970.

Fifty-three women undergoing hysterotomy for psychiatric reasons at a time between 13 and 24 weeks' gestation were examined to determine the accuracy of ultrasonic measurement of the fetal biparietal diameter during the second trimester of pregnancy. The mean and standard deviation of each of 3 ultrasonic measurements before delivery and external caliper measurements directly after delivery were small, indicating a high degree of reproducibility. The high degree of positive correlation between the ultrasonic and caliper recordings was highly significant statistically. The failure rate in performing cephalometry during the second trimester was 6%. The results demonstrate that the fetal head can be measured with a high degree of accuracy from the thirteenth week of pregnancy onwards by using a combination of 2 scan techniques. (12 refs.) - *B. J. Grylack.*

Queen Charlotte's Hospital
London W. 6, England

1579 **CHAMBERLAIN, GEOFFREY; & FOSKER, A. P.** Further simplifications in the detection of fetal haemoglobin in clinical practice. *Journal of Obstetrics and Gynaecology of the British Commonwealth*, 77(12):1096-1097, 1970.

A tablet containing sodium metasilicate, magnesium stearate, starch, and microcrystalline cellulose was prepared for usage in distinguishing between adult and fetal hemoglobin in small samples of blood. The tablets in the single tablet test were checked by utilizing 25 pairs of samples of known maternal venous and fetal bloods. In each case, clear end points served as a basis for differentiation. Blood coming from the vagina during pregnancy or labor is usually maternal in origin. However, it is sometimes fetal, and subsequent management is determined by the distinction. In addition to its usefulness in ruptured vasa previa, the tablet is valuable in fetal blood sampling procedures, amniocentesis, and when blood is swallowed by the baby at delivery. (4 refs.) - *B. J. Grylack.*

Queen Charlotte's Hospital
London W. 6, England

1580 **MONKUS, ELLEN ST. J.; NYHAN, WILLIAM L.; FOGEL, BERNARD J.; & YANKOW, SANFORD.** Concentrations of uric acid in the serum of neonatal infants and their mothers. *American Journal of Obstetrics and Gynecology*, 108(1):91-97, 1970.

Testing for disorders of purine metabolism in infants should be done after 3 days of life, and elevated concentrations of urate observed during the first 2 days should probably be followed up with additional determinations after 3 days. Infant serum concentrations of uric acid measured in 45 normal mother-infant pairs were correlated with those of the mother up to 48 hours after birth and did not begin to reflect what was occurring in the infant until after 72 hours. Diagnosis of hyperuricemia related to the Lesch-Nyhan and other syndromes in families at risk can be made definitely by assay of the enzyme hypoxanthine-guanine phosphoribosyl transferase in the erythrocyte as early as the first day of life. (12 refs.) - *J. K. Wyatt.*

University of California, San Diego
La Jolla, California 92037

1581 **BURGESS, MICHAEL M.; KODANAZ, ALTAN; & ZIEGLER, DEWEY K.** Prediction of brain damage in a neurological population with cerebrovascular accidents. *Perceptual and Motor Skills*, 31(2):595-601, 1970.

Fifty-four brain-damaged patients and 21 patients without brain damage who had been hospitalized because of possible neurological effects of a documented cerebrovascular accident were given a test battery containing 15 intellectual and 12 sensory-motor variables. The object of testing was to determine whether intellectual and sensory-motor measures significantly differentiate brain-damaged from non-brain-damaged cerebrovascular accident patients, whether multiple variant prediction improves brain-damage diagnosis in the given clinical population, and whether the performance on tests by brain-damaged cerebrovascular accident patients is similar to that previously found in a spectrum of neurological and psychiatric patients. It was concluded from the results that sensory-motor measures are clearly superior to intellectual measures as single predictor variables, that brain damage which is measured behaviorally is consistent across patient populations in neurological, psychiatric, and neurological subgroups with cerebrovascular accident, and that multivariant prediction is a promising method for the diagnosis of brain damage in a cerebrovascular accident population. (15 refs.) - *B. J. Grylack.*

University of Missouri
Kansas City, Missouri

1582 **KOHN, GERTRUDE; & ROBINSON, ARTHUR.** Tetraploidy in cells cultured from amniotic fluid. *Lancet*, 2(7676):778-779, 1970. (Letter)

Three women who underwent amniocenteses were found to have tetraploidy and tetraploid mosaicism in cells cultured from amniotic fluid. A 29-year-old woman, mother of 2 sons with trisomy 21, had cells taken during the fourth month which revealed only tetraploid cells after 42 days of culture. A normal, full term female was delivered. A 14-year-old girl, examined during the twenty-sixth to thirtieth week because of suspected fetal malformation, revealed 13% tetraploid metaphases after 20 days of culture, none after 35 days, and delivered a full term normal male. A 28-year-old woman with one trisomy 21 male offspring was examined during the fourth month of gestation and had 56% of tetraploid cells after 14 days of culture. New amniotic fluid taken 3.5 weeks later showed 50% tetraploid cells after 11 days of culture. The pregnancy was terminated intentionally. The discovery of a significant number of tetraploid cells in amniotic fluid cultures necessitates the determination of whether the finding constitutes a normal phenomenon or an indication for termination of pregnancy. (2 refs.) - *B. J. Grylack.*

Children's Hospital
Philadelphia, Pennsylvania 19146

1583 **WAHLSTROM, J.; BROSSET, A.; & BARTSCH, F.** Viability of amniotic cells at different stages of gestation. *Lancet*, 2(7681):1037, 1970. (Letter)

The best time for amniocentesis for chromosome analysis is between weeks 15 and 18 of gestation.

This permits an optimal balance between culturing conditions and abortion. The concentration of living amniotic cells seems to increase between weeks 11 and 34. These conclusions are based on a study of 26 specimens of amniotic fluid obtained from legal abortions and amniocentesis because of Rh immunization problems. (3 refs.) - *E. Kravitz.*

St. Jorgen Hospital
Hisings Baika, Sweden

1584 **ROUX, J. F.; WILSON, R.; YENIKOMSHIAN, H.; JASSANI, M.; & JORDAN, J.** Labor monitoring: A practical experience. *Obstetrics and Gynecology,* 36(6):875-880, 1970.

Experiences with electronic monitoring of labor, e.g., intrauterine pressure, fetal heart rate, and scalp pH, are described. Introduction of the monitoring devices caused major discomfort in 25% of the patients and interfered with patient care. There were patient or technical difficulties in half the cases. Although about a fourth of the patients were frightened, nearly all liked the medical attention represented by the monitoring. Electronic monitoring requires a 24-hour watch by professional personnel and may not be too practical on a routine basis in a community hospital. This procedure might best be limited to selective cases. (12 refs.) - *E. Kravitz.*

Case Western Reserve University
School of Medicine
Cleveland, Ohio

1585 Ultrasound: 'Safe for the fetus'. *Medical World News,* 12(8):5, 1971.

Sonography is considered safe for a human fetus because it directs intermittent impulses of mechanical energy, rather than ionizing radiation, to a point on the body. In a survey of 1,124 normally pregnant women in whom sonography was utilized to diagnose early pregnancy, to aid in localization of the placenta, or to study fetal development, the 2.7% abnormality rate was reported to be lower than the average of the general population. Although skepticism still exists among some physicians, the bulk of the evidence appears to favor the use of this technique. (No refs.) - *B. J. Grylack.*

1586 **MURPHY, ANN; & POUNDS, LOIS.** Repeat evaluations of retarded children. *American Journal of Orthopsychiatry,* 42(1):103-109, 1971.

Qualitative assessment of 58 families with MR children who sought additional diagnosis from a university mental retardation unit suggested 6 categories of parental motivation for repeat evaluation: (1) search for a treatable organic diagnosis rather than an emotional disorder; (2) attempt to obtain professional support that no defect existed; (3) search for guidelines for planning required at a new developmental stage; (4) those referred by physicians who did not wish to deal with a retarded patient; (5) those referred by social agencies who were using the evaluation as a way of postponing a difficult decision; and (6) search for a more positive response from professional people, particularly in the area of remedial measures that might enhance the child's development. The last 4 categories represent inability of professionals to meet the family's needs. Suggestions for better professional service include (1) exchange of information with referral sources regarding additional data on the child's handicap that would facilitate planning; (2) initial interviews with parents who are suspected of denying the diagnosis to identify how a repeat evaluation will serve the client; and (3) a more active and positive response from professional people to foster better clinic-family interaction with parents of the profoundly retarded. (7 refs.) - *J. C. Moody.*

Children's Hospital Medical Center
Boston, Massachusetts 02115

1587 **FEINGOLD, MURRAY; & GELLIS, SYDNEY S.** Syndrome Identification and Consultation Service. *American Journal of Diseases of Children,* 121(1):82-83, 1971.

A Syndrome Identification and Consultation Service, funded by the Massachusetts Bay Chapter of the National Foundation – March of Dimes, has been moderately successful in the diagnosis of the unusual looking child. Mail requests for diagnosis were submitted from all areas of the United States and from many foreign countries. A group of 10 consultants was able to make a definite diagnosis in 18% of the first 100 cases. Where a definite diagnosis was not made, the consultants entertained some type of diagnosis in 35% of the cases. Diagnoses

utilized color slides (although black and white photographs would assure better quality), roentgenograms, and an atlas or textbook photographs in addition to history, physical, and laboratory findings. Until improved methods such as closed circuit television or computer assisted diagnosis are feasible, the service warrants continued use as a diagnostic aid. (7 refs.) - *S. Glinsky.*

Boston Floating Hospital for Infants and Children
Boston, Massachusetts 02111

1588 **OSOFSKY, HOWARD J.; LONG, ROBERT E.; O'CONNELL, EDWARD J., JR.; & MARSHALL, LINDA D.** Estrogen excretion during pregnancy in a high-risk population. *American Journal of Obstetrics and Gynecology*, 109(1):1-7, 1971.

Twelve hundred and ninety-two urine specimens were collected from 219 consecutive low-income pregnant adolescent girls to determine estrogen/creatinine ratios during pregnancy and the value of that ratio as a clinical adjunct in high-risk pregnancies. The absence of any extremely low values beyond the thirty-fourth week of gestation in the sample seems to have prognosticated the favorable outcome for all of the girls correctly. The mean estrogen/creatinine ratios correlated highly significantly with infant birth weight ($p<0.01$). Two other maternal parameters, weight gain during pregnancy and number of weeks of gestation at delivery, also correlated with infant birth weight ($p<0.01$). The results suggest that within the range of normal estrogen excretion patterns it may be possible to predict abnormally small infant size. The absence of correlation between mean estrogen values from the thirtieth week of gestation to delivery, and week of gestation at delivery and maternal weight gain during pregnancy, the 2 other factors related to infant birth weight, suggests a specific and different hormonal relationship to the area of investigation. (22 refs.) - *B. J. Grylack.*

State University of New York
Syracuse, New York

1589 **KRASNER, JOSEPH; JUCHAU, MONT R.; NISWANDER, KENNETH R.; & YAFFE, SUMNER J.** Theoretical considerations regarding the presence of biliverdin in amniotic fluid. *American Journal of Obstetrics and Gynecology*, 109(1):159-165, 1971.

To explain the presence of biliverdin pigment in early pregnancy, human amniotic fluid obtained between the eighth and sixteenth weeks of gestation was analyzed for biliverdin, and human biliverdin reductase activity in the fetal-placental unit was measured at several gestational stages. The presence of the pigment was confirmed by column chromatography, electrophoresis, ultraviolet fluorescence, thin-layer chromatography, and spectral analysis. The results indicated the presence of biliverdin in human amniotic fluid during early pregnancy and the lack of fetal capacity to reduce the pigment enzymatically. The concentration of biliverdin in amniotic fluid probably reflects an interplay of several factors, including hemoglobin synthesis, conversion of hemoglobin to biliverdin, biliverdin reduction to bilirubin, and transfer into the maternal circulation. It appears likely that the ultimate source is fetal erythropoietic tissue and that entry into amniotic fluid would follow the normal fetal excretory routes. (15 refs.) - *B. J. Grylack.*

Children's Hospital
Buffalo, New York 14222

1590 **CURL, CHARLES W.** Immunoglobulin levels in amniotic fluid. *American Journal of Obstetrics and Gynecology*, 109(3):408-410, 1971.

Amniotic fluid from 10 term obstetric patients was examined for immunoglobulins. The fluid was obtained by transvaginal needle aspiration through intact fetal membranes, and the detection of immunoglobulins was achieved by immunodiffusion and electroimmunodiffusion techniques. The detection of immunoglobulin (Ig) A in the amniotic fluid of 8 out of 10 patients is significant, although the actual immune activity of this immunoglobulin has not been confirmed. IgG and IgM were not found in amniotic fluid in amounts detectable by the employed method. The detection of immunoglobulins in amniotic fluid has potential value, for it may represent a means of prenatal detection of intrauterine infection which would allow planning for or execution of diagnostic or therapeutic measures. (34 refs.) - *B. J. Grylack.*

University of Washington
Seattle, Washington

1591 **GLUCK, LOUIS; KULOVICH, MARIE V.; BORER, ROBERT C., JR.; BRENNER, PAUL H.; ANDERSON, GERALD G.; & SPELLACY, WILLIAM N.** Diagnosis of the respiratory distress syndrome by amniocentesis. *American Journal of Obstetrics and Gynecology*, 109(3):440-445, 1971.

Two hundred and seventy-two pregnancies were studied with 302 amniocenteses to determine the usefulness of phospholipid evaluation in correlation with pulmonary maturity of the fetus in predicting the likelihood of the respiratory distress syndrome in a premature birth. During gestation, total lipids increased in concentration and rose suddenly at 35 weeks. On the basis of thin-layer chromatography, the nonacidic phospholipid fraction, which consists mainly of sphingomyelin and lecithin, was found to be the most important and showed a sharp spurt in concentration at 35 weeks. The relationship between these 2 compounds is the basis for clinical evaluation of lung maturity and allows for determination of a date after which the respiratory distress syndrome would be unlikely if the child is born. Surface-active amniotic fluid lecithin at birth and lecithin from tracheal effluent taken immediately after birth were very similar in fatty acid composition and were uniquely saturated, characteristic of the kinds of lecithin found only in the lung. (22 refs.) - *B. J. Grylack.*

University of California,
San Diego School of Medicine
La Jolla, California

1592 **GOLDSTEIN, ROBERT; & TAIT, CHARLES.** Critique of neonatal hearing evaluation. *Journal of Speech and Hearing Disorders*, 36(1):3-18, 1971.

The most popular approach to neonatal hearing screening is to screen all newborn infants in the nursery prior to discharge from the hospital, the preferred test signal being a 3,000-Hz warble tone with a 90 dB SPL screening level and with grading on a 5-point scale. Positive features of a routine neonatal hearing screening program include the presence of almost all newborns in 1 place for mass screening, the opportunity to detect deafness in a baby not known to be high-risk and thereby to alert the physician to a more widespread congenital disorder, and a stimulus for professionals to know more about auditory ability. Negative features center around the magnitude of the problem, the effectiveness of the screening procedure, and the significance of follow-up procedures. Despite needed progress in the accuracy and organization of testing, the concepts of early detection of hearing impairment and early remedial measures including amplification are supported. (11 refs.) - *B. J. Grylack.*

University of Wisconsin
Madison, Wisconsin

1593 A test for glycogen disorder. *Medical World News*, 12(9):31, 1971.

Brancher deficiency, or amylopectinosis, and Pompe's disease can now be identified by culturing cells from amniotic fluid at an early enough stage in fetal development to allow a therapeutic abortion. Of the 8 currently known glycogen storage diseases, these 2 rare diseases are untreatable and lethal early in life. Amniotic fluid specimens are obtained between the fourteenth and sixteenth weeks of pregnancy, and results are usually confirmed by the twentieth week of gestation, when it is still safe for abortion. In Pompe's disease, clinical signs include profound hypotonia, a massively enlarged heart, and cardiac failure without cyanosis during the first year of life. In brancher deficiency, although the patient appears normal at birth he soon manifests an enlarged and progressively cirrhotic liver. At the present time, the test and the amniocentesis procedure are prohibitively expensive. Since the diseases are inherited in an autosomal recessive manner, plans are being made to organize familial studies on a national scale. (No refs.) - *B. J. Grylack.*

1594 **GERBIE, ALBERT B.; NADLER, HENRY L.; & GERBIE, MELVIN V.** Amniocentesis in genetic counseling. *American Journal of Obstetrics and Gynecology*, 109(5):765-768; discussion, 768-770, 1971.

Two hundred and fifty-six transabdominal amniocenteses were performed between the thirteenth and eighteenth weeks of fetal gestation in 231

outpatients during 238 pregnancies in which there was significant risk that the child would be born with serious genetic defects. There was no maternal morbidity, hemorrhage, infection, or death. Neither a spontaneous late abortion occurring 1 month after amniocentesis in 1 patient nor a fetal death at term in another patient was attributable to the procedure. Five sets of similarly sexed twins were delivered. The results of amniotic fluid analysis predicted the fetal genetic status correctly in all but 2 instances, in which the overgrowth of maternal cells in culture probably interfered. Transabdominal amniocentesis performed early in the second trimester of pregnancy carries minimal risks to mother and fetus. However, the ability to detect a genetic defect in the fetus has created many moral, legal, and medical problems. Effective treatment of genetic defects is limited at the present time, and therapeutic abortion is legal in relatively few states. While the value of selecting generally high-risk pregnancies for amniocentesis for cytologic, cytogenetic, and enzymatic analyses is disputed, screening of women known to have transmissible chromosomal anomalies is clearly indicated in many cases. (21 refs.) - *B. J. Grylack.*

Northwestern University Medical School
Chicago, Illinois

1595 SLATER, STANLEY. An evaluation of the metacarpal sign (short fourth metacarpal). *Pediatrics*, 46(3):468-471, 1970.

In a study undertaken to determine the clinical prevalence of short fourth metacarpal bones in a sample of the general population, 9.6% of an unselected group of 1,510 patients over the age of 6 years had short fourth metacarpals. X-ray examination of several of the Ss disclosed 10% false positives. Maximum shortening was 3 mm clinically and 2.5 mm on X-ray. This approach toward quantitation of the degree of shortening should provide for better comparison between apparently normal Ss with shortened fourth metacarpals and patients with ovarian agenesis, pseudohypoparathyroidism, and pseudopseudohypoparathyroidism. Shortening in excess of that found in normal Ss may provide the clue to the possible existence of such abnormalities and indicate the need for endocrine evaluation. (6 refs.) - *M. S. Fish.*

Bay Ridge Medical Building
Brooklyn, New York 11220

1596 HAFFTER, C. Zukunftsperspektiven der pranatalen padiatrie (Future perspectives in prenatal pediatrics). *Acta Paedopsychiatrica*, 37(910):241-243, 1970. (Editorial)

High risk of severe malformations in the child has been for some time a 'eugenic' indication for interruption of pregnancy, especially in the Scandinavian countries. The accuracy of prediction has been greatly improved through amniocentesis and the subsequent cell investigation, which allows an accurate diagnosis to be made *in utero*. Consequently, therapeutic abortion is possible before the 18th week. Investigation is indicated if the mother is known to be a carrier of a D/G translocation or enzyme defects such as galactosemia or homocystinuria. A change in other circumstances—the ethical attitudes of physicians and jurists, improved possibilities for prediction of genetic fetal damage, the demand by the mother of the right to part with a fetus of unwanted sex—might eventually lead to complete freedom for the interruption of pregnancy. (No refs.) - *P. Custers.*

Schaffhauser Rheinweg 55, CH
4058 Basel, Switzerland

1597 BARAKAT, BASSAM Y.; AZOURY, RAMEZ S.; & JONES, HOWARD W., JR. Determination of fetal sex during the second trimester of pregnancy. *Obstetrics and Gynecology*, 37(1):134-140, 1971.

Sex chromatin analysis is a reliable method for determining fetal sex during the second trimester of pregnancy. The technic employs amniotic fluid cells grown in short-term culture. Amniotic fluid specimens were obtained from 28 patients between week 12 and week 20 of gestation. In chromatin-positive specimens counts were slightly lower than for normal buccal smears in the laboratory and ranged from 11 to 28% for initial determinations in culture. In 23 specimens from 22 cases of therapeutic abortion, sex chromatin and fetal gonadal sex correlated completely. In 6 patients studied for the possible presence of a genetically defective fetus, there was full agreement between sex chromatin and karyotype from amniotic fluid culture. This test in conjunction with antenatal chromosome analysis will lead to

a reliable cytogenetic diagnosis in the investigation of high-risk pregnancies. (19 refs.) - *S. Glinsky.*

Johns Hopkins University School of Medicine
Baltimore, Maryland 21205

1598 **BRUKKER, BRUCE H.; REYNOLDS, WILLIAM A.; ORTIZ, EDUARDO; & JOSEY, JULIAN C.** Placental localization. *American Journal of Obstetrics and Gynecology,* 110(1):9-14, 1971.

For placental localization isotopic placentography is preferable to amniography, which should be used only if isotopic placentography is unavailable. Both radioiodinated serum albumin (RISA) placentography and amniography were done for placental localization in an unselected series of 28 patients with third-trimester vaginal bleeding. Radiation to the fetus in isotopic placentography is considerably less than that required by amniography. Further, results indicate that isotopic placentography is more accurate, with an accuracy rate of 97% as opposed to 86% for amniography, is simpler, and avoids the potential hazards of amniocentesis. While amniography is a useful diagnostic tool, it should not be used when isotopic placentography is available for placental localization. (16 refs.) - *S. Glinsky.*

Henry Ford Hospital
Detroit, Michigan

1599 **FREEMAN, ROGER D.** A child psychiatrist looks at mental retardation. *Deficience Mentale/Mental Retardation,* 21(1):11-14, 1971.

Due to the tendency to categorize all MRs as 1 group, the complexity of diagnosis in early childhood, and the confusing nature of terminology, the comprehension of MR is often difficult. The psychiatrist must know the limitations of early diagnosis and must be cautious when he advises parents. Early intervention is advisable in many cases, but it may be difficult to determine when and how to intervene. The professional staff must give continuous support to parents and should aid them in interpreting the situation for their retarded child. There is no agreement concerning the percentage of MRs with behavioral disturbances. It is suspected that much of the behavioral and emotional difficulty encountered by the retardate is related to repeated frustrating experiences, to a lack of success needed to build self-esteem, and to the tensions which may develop with parents and others during the child's early years. (No refs.) - *B. J. Grylack.*

University of British Columbia
Vancouver, British Columbia, Canada

1600 **WILLIAMS, BARBARA L.; & RICHARDS, SHARON F.** Fetal monitoring during labor. *American Journal of Nursing,* 70(11):2384-2388, 1970.

The value of fetal monitoring and the procedures employed at the University of Washington Medical Center, where monitoring is used routinely during the first 2 stages of labor, are discussed. Fetal scalp ECG and intrauterine pressure are measured with a Corometrics unit; fetal heart sounds and abdominal contour changes are measured by the Cardiotocograph. The Doppler ultrasonic motion sensor (Doptone) is also used. The nurse can help to minimize patient discomfort and concern about the strange monitoring equipment. Graphic recorders are preferable to audible ones. (12 refs.) - *E. Kravitz.*

University of Washington
Seattle, Washington

1601 Dermatoglyphics. *Medical World News,* 11(31):24-29, 1970.

Dermatoglyphics, the study of prints of palms of hands and soles of feet, is discussed as a possible aid in the diagnosis of certain pathologies. This method is particularly useful in the case of mongolism or rubella embryopathy. It also seems useful in some chromosomal aberrations, crib death, congenital heart disease, leukemia, and possibly other malignancies. Although statistically significant differences in prints have been found in certain conditions, clinical diagnostic applicability to a single patient has been limited because of a wide range for normal values. The technique is simple to use, inexpensive, and painless, but considerable research is still needed. (No refs.) - *E. Kravitz.*

1602 **ALBERMAN, EVA; & GOLDSTEIN, HARVEY.** The at-risk register. *Lancet*, 2(7675):726-727, 1970. (Letter)

The testing of a high-risk group of children for a particular condition, by concentrating available resources on this group, is suggested as an improvement over the at-risk register. Such an approach is more economical per unit of abnormality discovered, and it would utilize limited resources more realistically. The present method of universal screening of children for handicapping conditions not detectable at birth has not been too effective. (2 refs.) - *E. Kravitz.*

National Bureau for Co-operation in Child Care
London, W. 1., England

1603 **HALLIDAY, N. P.** Assessment of gestational ages of neonates. *Nursing Mirror*, 131(19):26-30, 1970.

A semiquantitative method is provided for the estimation of the approximate gestational age of a newborn infant. This procedure distinguishes between premature and full-term neonates; it also distinguishes normal full-term from dysmature infants. The latter decision is aided by noting the birth weight on a prepared chart. The dysmature infant is particularly vulnerable to hypoglycemia. Various tests and criteria are given to aid the midwife in screening the neonates, e.g., Moro reflex, wrist and ankle flexion, scarf sign, heel-to-ear maneuver, plantar creases, posture, and head rotation. (No refs.) - *E. Kravitz.*

King's College Hospital
London, England

1604 **JENKINS, G.H.C.** At-risk register. *Lancet*, 2(7677):822, 1970. (Letter)

It is suggested that assessment by physicians is preferable to an at-risk register for children who might need regular care for developmental defects. One physician, with proper help, can manage the children of a community of 20,000 people. Virtually 100% immunization can be achieved. The child's health is studied as part of a family evaluation. (2 refs.) - *E. Kravitz.*

Ashford
Middlesex, England

1605 **MINCHIN, A. J.; & BHOYROO, S. K.** Feto-maternal transfusion in antepartum haemorrhage, an index of placental damage and fetal risk. *Journal of Obstetrics and Gynaecology of the British Commonwealth*, 77(9):781-784, 1970.

Seven obstetrical patients with uterine bleeding between 18 and 27 weeks of maturity and 29 cases of antepartum hemorrhage were studied to determine whether a relationship exists between the degree of feto-maternal transfusion and the fetal outcome. A minimum of 500,000 fetal erythrocytes were counted in the blood taken within 24 hours of the hemorrhage. In the cases of bleeding before 28 weeks, a high fetal cell count had a poor prognosis of either abortion or stillborn delivery. It was found that the greater the feto-maternal transfusion, the greater was the incidence of placental infarction and of fetal distress in labor. The fetal cell count may prove to be a useful index for predicting fetal risk and for determining subsequent management of the pregnancy. (10 refs.) - *B. J. Grylack.*

Hammersmith Hospital
London, W.12, England

1606 **ROOPNARINESINGH, SYAM.** Amniotic fluid creatinine in normal and abnormal pregnancies. *Journal of Obstetrics and Gynaecology of the British Commonwealth*, 77(9):785-790, 1970.

Specimens of amniotic fluid were obtained from obstetrical patients at the time of elective Cesarean section, hysterotomy, amniocentesis for rhesus isoimmunization, and artificial rupture of membranes for induction and acceleration of labor and were evaluated for creatinine concentration of the fluid in normal and abnormal pregnancies at various periods of gestation. There were 99 determinations from uncomplicated pregnancies, 5 from patients with diabetes mellitus, 1 from an eclamptic, and 25 from mothers with pre-eclamptic toxemia. The results showed a consistent linear increase in creatinine as pregnancy progressed, with variations among patients at different stages. In toxemia there was a linear rise with the same rate of increase, although absolute levels of creatinine were higher than in normals. A significant correlation between birthweight and creatinine values was also noted. As the fetus matures, creatinine appears to be found in increasing amounts in the amniotic fluid because it is filtered completely by the

increasing number of functional glomeruli and is not reabsorbed by the tubules. Findings indicate that estimates of creatinine level in amniotic liquor in normal patients may be useful in distinguishing between the premature and dysmature infant and in estimating fetal maturity. (14 refs.) - *B. J. Grylack.*

University of the West Indies
Jamaica, West Indies

1607 **O'NEILL, ROBERT T.; MORROW, GRANT, III; HAMMEL, DEBORAH; AUERBACH, VICTOR H.; & BARNESS, LEWIS A.** Diagnostic significance of amniotic fluid amino acids. *Obstetrics and Gynecology*, 37(4):550-554, 1971.

Investigation of amino acid concentrations in 27 specimens of amniotic fluid of 8 to 44 weeks gestation indicated that all amino acid concentrations decreased during gestation; ethanolamine and taurine increased. Comparison with 3 first-voided newborn urines suggested that the contribution of fetal urine toward the total concentration of amino acid in amniotic fluid is minor. A normal amniotic fluid-amino acid pattern was found in a fetus at term with maple syrup urine disease. The value of prenatal quantitation of amniotic fluid amino acids in the detection of fetal aminoacidopathies is doubtful since maternal amniotic fluid concentrations remain normal unless the mother is affected with a disorder of amino acid metabolism. (21 refs.) - *J. K. Wyatt.*

Hospital of the University of Pennsylvania
Philadelphia, Pennsylvania 19104

1608 **GLUCK, L.; KULOVICH, M. V.; BORER, R. C.; BRENNER, P. H.; ANDERSON, G. G.; & SPELLACY, W. N.** Accurate prediction of lung maturity and respiratory distress syndrome (RDS) by amniocentesis. *Obstetrics and Gynecology*, 37(4):631, 1971.

Amniotic fluid examination can be used to assess fetal lung maturity since the development of the lung is reflected by lipid changes in the fluid. Until the thirty-fifth week of gestation, lecithin and sphingomyelin concentrations in the amniotic fluid do not differ markedly. At week 35, a sharp increase in the concentration of surface lecithin takes place. This surge in lecithin marks lung maturity and occurs because of synthesis by one of the 2 pathways for lecithin biosynthesis (CDP-choline+diglyceride). Respiratory distress syndrome will not develop if the infant is delivered at this time. (No refs.) - *J. K. Wyatt.*

University of California
San Diego, California

1609 **BOLOGNESE, RONALD J.; CORSON, STEPHEN L.; TOUCHSTONE, JOSEPH C.; & LAKOFF, KENNETH M.** Correlation of amniotic fluid estriol with fetal age and well-being. *Obstetrics and Gynecology*, 37(3):437-442, 1971.

Amniotic fluid estriol concentration may be a more practical and more meaningful measurement of fetal status and gestational age than the 24-hour total urinary value. Fifty-seven amniotic fluid samples were obtained from 52 patients (gestation range 16 to 43 wks), and 24-hour urinary estriol was determined concomitantly in 20 cases. Changes in amniotic fluid estriol over a 5-week period were slight for 25 patients between 16 and 21 weeks gestation who were undergoing saline-induced pregnancy termination. As pregnancy progressed, 27 patients between 26 and 43 weeks gestation had significantly ($p<.001$) higher concentrations of estriol in amniotic fluids. A close linear relationship between simultaneous urinary and amniotic fluid determinations on 20 patients could not be established, although the correlation was generally in a positive direction. (13 refs.) - *J. K. Wyatt.*

829 Spruce Street
Philadelphia, Pennsylvania 19107

1610 **CASAER, P.; & AKIYAMA, Y.** Neurological criteria for the estimation of the post-menstrual age of newborn infants. In: Stoelinga, G.B.A.; & Ven Der Werff Ten Bosch, J. J., eds. *Normal and Abnormal Development of Brain and Behaviour.* Baltimore, The Williams & Wilkins Co., 1971, p. 148-175.

Neonatal mortality depends more on post-menstrual (PM) age than on birthweight, since

physiologic functions may depend strongly on such age. For developmental tests, PM age is more relevant than age since birth. Nonneurological parameters—body and bone growth, skin and heart changes, and physicochemical changes—are useful, but most important are the developmental neurological parameters: (1) Brain/skull growth (biparietal diameter of skull and postnatal head circumference); (2) Neuromotor function (prenatal quickening and postnatal neurologic tests and polygraphic data); (3) Electrophysiologic data (postnatal nerve-conduction velocity, EEG, and evoked response); (4) Visual system (postnatal eye fundus, electroretinography, and photically evoked responses). Neurologic tests include postural responses. Polygraphic studies involve measurements of sleep-cycle characteristics. Events that appear abruptly at particular ages are ideal for estimating PM age. (112 refs.) - *B. Berman.*

1611 **SCHIFFER, MORTON A.; ERTAL, NORMAN A.; HELLMAN, LOUIS M.; & KOBAYASHI, MITSUO.** Combined method for evaluating fetal well-being by plasma estriol measurements and ultrasonography. *American Journal of Obstetrics and Gynecology*, 108(8):1277-1286, 1970.

The simultaneous use of estriol levels and ultrasonography accurately assesses the fetal condition of high-risk patients who have had multiple plasma estriol measurements taken and 1 or more ultrasonograms made. Estriol measurements were of value in estimating the fetal condition, if the fetal size and age were known. Combined use of plasma estriol measurements and ultrasonographic cephalometry resulted in a more accurate assessment of fetal well-being, size, and growth and a great reduction in the risk of prematurity. Further combined studies are needed to clarify the relative values of estriol levels and ultrasonography in the estimation of fetal status. The criteria of normal fetal growth and welfare in high-risk situations must be established to decrease high perinatal loss rates. (10 refs.) - *S. Glinsky.*

The Jewish Hospital
Brooklyn, New York

1612 **SCHULTE, F. J.** Neonatal EEG and brain maturation. *Developmental Medicine and Child Neurology*, 12(5):681-682, 1970.

Objections by another investigator to the author's study on neonatal EEG and brain maturation are based on incorrect citations and misunderstandings. The other investigator's data are based by his own words on infants who reached term, and the author and his colleagues did not define maturational changes in the EEG without specific reference to sleep states, but within each state. Also, at 35 weeks conceptional age there are different states of sleep and very different EEG patterns recurring in a particular sequence together with certain behavioral characteristics. Interpreters will not be able to make very many correct evaluations of the EEGs and infant maturity when presented with only the EEGs of pre-term and full-term newborn infants, both normal and abnormal. The author and his colleagues found it necessary to record at least 1 total sleep cycle and to include channels for eye movements, respiration, and muscle activity. Knowledge of the behavioral state was mandatory to make correct evaluations of infant maturity high above a level of significance. (8 refs.) - *S. Glinsky.*

Universitaets-Kinderklinik
34 Goettingen, West Germany

1613 **WOOD, C.B.S.** The development of immunoglobulins. *Developmental Medicine and Child Neurology*, 12(5):663-666, 1970.

Increasing knowledge of the ontogenesis of immunological functions in fetal and infant life, particularly of the immunoglobulins IgG, IgA, IgM, IgD, and IgE, has important implications in the detection of congenital infections and for active immunization prophylaxis. IgM, IgA, and specific IgM antibodies, which can be synthesized by the fetus *in utero*, have been reported in a number of congenital infections, including congenital rubella, cytomegalic infection, toxoplasmosis, and herpes simplex. But the use of high levels of IgM in cord sera as a diagnostic criterion of intrauterine infection is open to question, since a recent study showed that IgM concentration may be raised in only 20% of cases of fetal rubella infection. IgG, which is passively acquired from the mother, seems to be directly related to gestational age, but it is doubtful whether immunoglobulin should be given routinely to short gestation infants, since injection of the partially purified preparations at present available

may induce isoimmunization in immunologically competent recipients. (30 refs.) - *S. Glinsky.*

Royal Hospital for Sick Children
Bristol, England

1614 NELLHAUS, GERHARD. Cerebrospinal fluid immunoglobulin G in childhood. *Archives of Neurology,* 24(5):441-448, 1971.

In children the upper limit of normal cerebrospinal fluid (CSF) immunoglobulin G (IgG) measured by electroimmunodiffusion (EID) is 8.2%, with higher values suggesting demyelinating processes. The Ss were 160 children from 1 through 15 years of age and 73 control children aged 1 to 14 years old. In children with nondemyelinative neurologic disorders, the mean CSF IgG quotient of protein was 4.6%, compared to 7.1% in "normal" adults in the same laboratory, a highly significant difference. In 88% of the controls, CSF IgG was below 6.4%, and in 99%, 8.2% or less. In 88% of the children with "demyelinative" diseases, exclusive of subacute sclerosing panencephalitis, CSF IgG was 8.3% or higher, but significantly lower than the 14% considered compatible with multiple sclerosis in adults. The method of EID requires only minute amounts of CSF and is recommended for wider clinical use in children. (22 refs.) - *S. Glinsky.*

University of Colorado Medical Center
Denver, Colorado

1615 CASSADY, GEORGE; & BARNETT, ROBERT. The predictive value of amniotic fluid spectrophotometry. *American Journal of Obstetrics and Gynecology,* 108(7):1010-1016, 1970.

Amniotic fluid spectrophotometry is the most accurate tool available in the management of the Rh(D) sensitized pregnancy. Over a period of 4½ years, 685 amniocenteses were performed in 367 pregnancies. In 234 fluids from 178 pregnancies in Rh(D) positive women, amniotic fluid bilirubin was surprisingly elevated. In 451 fluids from 189 pregnancies in Rh(D) negative women, maternal Rh(D) sensitization was diagnosed. These data indicate the importance of amniocentesis at low titer and the value of frequent serial examinations, and reinforce the conclusion that amniotic fluid bilirubin content at time of delivery accurately reflects fetal condition. The standard Liley method is clearly superior in estimating amniotic fluid bilirubin content. (31 refs.) - *S. Glinsky.*

University of Alabama Medical Center
Birmingham, Alabama

1616 WARKANY, JOSEF. Syndromes. *American Journal of Diseases of Children,* 12(5):365-370, 1971.

The proliferation of syndromes of multiple congenital malformations serves no useful purpose unless consideration is given to the syndrome's etiology, pathogenesis, and prognostic value. Syndromes are classified according to different principles, such as anatomical anomalies, clinical findings, histologic or embryonic origin, chemical analysis, gene and chromosomal abnormalities, and environmental teratogenic agents. A syndrome fever seems to be spreading; both the naming of syndromes and their potential usefulness are debatable. At present an eponymic designation of syndromes, with restrictions, is perhaps most useful, since gaining a concise picture of the syndrome is the desired result. But the increasing reporting of new cases and syndromes is relatively unproductive, unless this stimulates etiologic research, statistical evaluation, or experimentation. The analysis and classification of multiple congenital malformations by means of recurrent associations of the disease is a step in the right direction. (32 refs.) - *S. Glinsky.*

Children's Hospital Research Foundation
Cincinnati, Ohio

1617 What tests should be routine for diagnosing and treating neurological disorders? *Expectations,* 1(1):1-2, 1971.

At present, there is no standard, complete battery of tests for determining the presence and extent of mental handicaps in young children. Many children are wrongly labeled as mentally handicapped when they actually have treatable metabolic or other conditions. Examinations and tests are often delayed, poorly chosen, badly

administered, and wrongly interpreted. More often than not, neurologically hurt children have a combination of disabilities, and treating one area pinpointed by too narrow a testing program does not constitute a real cure. A recommended program, including a chemical survey, allergy screening, and testing for inborn errors in utilization of amino acids, sugars, and B-complex vitamins, is presented. (No refs.) - *M. H. Street.*

1618 **MONIF, GILLES R. G.; & MENDENHALL, HIRAM W.** Immunoglobulin G levels and the titers of specific antiviral antibodies in amniotic fluid. *American Journal of Obstetrics and Gynecology*, 108(4):651-654, 1970.

Preliminary analysis of 43 amniotic fluid/maternal serum pairings obtained at the time of premature and term deliveries for complement-fixing antiviral antibodies and immunoglobulin G (IgG) levels determined by single radial diffusion technique indicates that specific antibody in amniotic fluid appears as early as 90 days gestation. The amount of specific antiviral antibody in the amniotic fluid is predominantly a function of the corresponding maternal titer and does not vary significantly during gestation. The amount of IgG detected in amniotic fluid at 25 to 34 weeks of pregnancy is quantitatively comparable to that observed in amniotic fluid samples obtained at term. Analysis of either specific antibody or IgG levels in amniotic fluid independently suggests that the distribution of maternally derived IgG and consequently specific antibody in amniotic fluid, unlike that in fetal circulation, does not change significantly with the progression of pregnancy. (10 refs.) - *J. C. Moody.*

University of Florida College of Medicine
Gainesville, Florida

MEDICAL ASPECTS – Prevention and Etiology (General)

1619 **REID, DUNCAN E.** The right and the responsibility. *American Journal of Obstetrics and Gynecology*, 108(5):825-832, 1970.

A study of 65,000 pregnancies with 12-year follow-ups of offspring is being designed to reveal quantitative risks of mental deficiency and neurological disorders in children from pregnancies complicated by a variety of medical diseases and unfavorable obstetrical conditions. There has been a 12.5% mortality rate in 1,500 to 3,000 gram infants at or near term. In general, uterine bleeding and infection appear to be the 2 most difficult factors to deal with obstetrically. In regard to MR without neurological signs, preliminary IQ screening at age 4 showed 47 per 1,000 with IQ<69. In the overlap of cerebral palsy with MR, the genetic role was associated with approximately 25%, approximately 50% were related to obstetric causes, and the remaining cases were not connected with any single factor. Attention should be given to fewer and larger services in order to provide obstetric and neonatal services that will significantly alter the results of high risk pregnancies. (No refs.) - *B. J. Grylack.*

No address

1620 **De GEORGE, FRANCES V.** Maternal and fetal disorders in pregnancies of mothers of twins. *American Journal of Obstetrics and Gynecology*, 108(6):975-978, 1970.

Pregnancy data on 329 mothers of twins over a 10-year period at a single medical center were studied to determine the relationship between various maternal and fetal conditions. Maternal and fetal disorders suggestive of abnormal placental development, including severe erythroblastosis fetalis, hereditary hemoglobinopathies, congenital nephrosis, congenital syphilis, hydatidiform mole, and hydropic infants from diabetic mothers, were evident in previous pregnancies in mothers of twins significantly more frequently than in control mothers of singletons. Hypospadias and enlarged clitoris were 2 anomalies that were also

found more often in previous offspring of twin-bearing mothers. The results of this and similar investigations indicate that certain maternal and pregnancy environmental conditions are associated with twinning, primarily dizygous, and the same conditions may also affect the reproductive performance of offspring. (21 refs.) - *B. J. Grylack.*

Upstate Medical Center
Syracuse, New York 13210

1621 Genetics. *Medical World News*, 11(38):6, 1970.

Data on chromosomal abnormalities among 4,500 babies born to women over age 34 in 1 year at the Yale-New Haven Hospital revealed a rate of 1.5% for such major defects as translocations, autosomal trisomies, and extra or missing sex units. In the U.S. in 1 year, 20,000 such babies are born. Amniocentesis is recommended for high-risk mothers, and if an odd karyotype is revealed therapeutic abortion should be considered. (No refs.) - *B. Berman.*

1622 CHASE, HELEN C.; & BYRNES, MARY E. Trends in "prematurity": United States, 1950-1967. *American Journal of Public Health*, 60(10):1967-1983, 1970.

The number of live births in the United States rose from 3.5 million in 1950 to 4.3 million in 1957. After a small decrease in 1958, the number increased again and reached a peak of 4.3 million in 1961. A steady decline followed until 1967, when the number of live births equaled that recorded for 1950. Observations on the nationwide trend in prematurity between 1950 and 1967 were made with regard to the percentage distribution of live births by period of gestation and color, by birth-weight and color, and by period of gestation within major birth-weight groups for registration areas in which the first day of the last menstrual period (LMP) was requested. Gestation age was then computed from the onset of this period to the date of birth in LMP areas. For non-LMP areas, gestation age was estimated by color and age of the mother. Observations were then made with regard to percent of preterm live births within specified birth-weight groups by color, proportion of plural births by color, percent change in live births by weight at birth and color, and associated ratios. The single most striking factor was the gradually increasing proportion of low-birth-weight infants for the nonwhite group simultaneous with a relatively stable rate for the white group. The improvement of gestation data and greater availability of socioeconomic indicators in the future will help to explain the significance of this factor more adequately. (12 refs.) - *B. J. Grylack.*

Association of Schools of Allied
Health Professions
Washington, D. C. 20036

1623 Any questions — uterine bleeding and fetal defects. *British Medical Journal*, 3(5714):91, 1970.

There is no firm evidence that uterine bleeding in pregnancy is a cause of congenital defects. A large proportion of spontaneous abortions show abnormal fetuses, so that malformations were the cause rather than the result of bleeding in these cases. Only about 5% of all pregnancies show threatened abortion. This is associated with a higher incidence of perinatal mortality but not with malformations. (6 refs.) - *E. L. Rowan.*

1624 Monitoring for malformations. *Nature*, 228(5270):415-416, 1970.

The problems associated with the study of congenital malformations were discussed at a symposium; mass screening seems impractical at present. Suggestions included the monitoring of spontaneous abortions to determine possible environmental influences at an earlier stage. Since specific major malformations are uncommon enough to require a wide range of surveillance, an alternative may be the observation for minor malformations. Japanese survivors of atomic radiation (exposed *in utero*) could be studied. Screening of medical records, chromosomes, and blood protein mutants may be difficult to do on a mass basis. (No refs.) - *E. Kravitz.*

1625 Oral feeding for the malnourished fetus? *Medical World News*, 11(36):13, 1970.

The possibility of feeding the fetus orally in cases of fetal hyponutrition has been examined by injecting ^{131}iodinated-(^{131}I) albumin into the amniotic cavity of 2 patients. Pregnancy was interrupted 20 hours later. On the basis of serial thyroid gammagrams, radioactivity curves in blood urine, feces from mother and child, chromosomal tests, and autoradiography of tissues, it was concluded that the fetuses swallowed, digested, and absorbed the injected compound. Albumin carrying the radioisotope appeared in the blood, thyroid, and urine of the newborn and also passed from the fetus to the mother. (No refs.) - *B. J. Grylack.*

1626 **BRADFORD, H. F.** Membrane potentials and metabolic performance in mammalian synaptosomes. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 1-11.

Neural cells often survive limited membrane penetration, probably because of cytoplasm viscosity that limits rate of loss of cofactors and enzymes or a resealing of the membrane. Thus, nerve-ending processes (much smaller regions of the neuron) are also able to survive disruptive forces, and–following axon and membrane rupture–form small, discrete, membrane-enclosed structures ("synaptosomes") that may be isolated by centrifugation. Their subsequent swelling and shrinking in varying media suggest resealing of the membranes. Demonstration of a potential across the synaptosomal membrane would be evidence of ion-accumulation transport function and metabolic integrity; this can be shown only by inference, since the synaptosome is too small for direct measurement. There is evidence that nerve endings retain much of the biochemical and morphological complexity they possessed *in situ*, thus providing opportunity for studying the relationship between synaptic events and metabolism. (28 refs.) - *B. Berman.*

Imperial College
London, England

1627 **McILWAIN, HENRY.** Distribution and utilisation of amino acids in the brain in relation to cerebral adaptation. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 15-23.

Among the causes of MR are amino-acid disturbances; consequently, it is important to study the distribution and metabolism of amino acids in brain tissues. Since MR, behaviorally, involves impairment of adaptation, it is important to understand the biochemical mechanisms involved in environmental adaptation, namely, the sequence from DNA to RNA to protein effectors. Involved in these mechanisms are the lability and responsiveness of cerebral proteins; the free amino acids which produce these proteins (these free amino acids undergo rapid turnover); and protein turnover. Excretion of valine and leucine reflects an important part of protein's normal amino-acid economy. Basic to MR is the mechanism by which the moving amino acids and the protein turnover are directed to produce lasting and biologically significant adaptation. Three considerations are important here: (1) a demonstration that protein synthesis was modified by stimulation of the brain or cerebral tissues; (2) in specifying the localization of metabolic change brought about by stimulation, transmitter release from nerve terminals is of outstanding importance; and (3) several agents involved in neurotransmission (serotonin and histamine) have been found to cause longer-lasting changes than these agents normally do. (21 refs.) - *B. Berman.*

University of London
London, England

1628 **BAERLOCHER, KURT; MOHYUDDIN, FAZL; & SCRIVER, CHARLES R.** Membrane transport of amino acids; use of mutation and ontogeny as probes of specificity. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 29-42.

Chemical, kinetic, and genetic studies of plasma-membrane permeability have demonstrated that the flow of organic solutes into and out of the cell is sustained by a network of proteins serving as transport sites. Influx and efflux permeabilities, as revealed by research into the ontogeny of the "iminoglycine" transport systems, develop independently, and the 3 physiological systems

for iminoacid and glycine influx appear to be independently controlled genetically. The phenotypic effects deriving from mutations that influence transport sites are consonant with the affected system and its significance for cellular nutrition. In certain gene-environment interactions, CNS growth and function are influenced by the inborn errors of amino-acid transport. (44 refs.) - *B. Berman.*

de Belle Laboratory for Biochemical Genetics
Montreal, Quebec, Canada

1629 **BORGAONKAR, DIGAMBER S.; & SHAH, SALEEM A.** Advances in human genetics and their impact on society. *Science*, 170(3955):347-348, 1970.

The nature of such scientific advances in human genetics as the detection of the chromosomal basis of Down's syndrome, the establishment of procedures for detecting genetic defects in the unborn child, and the clinical delineation of syndromes based upon data pertaining to the pattern of inheritance and the impact of these discoveries on social policies and programs requires that society become better informed about these matters and more involved in the decision-making process. One of the main reasons for seeking prenatal diagnosis is the consideration of therapeutic abortion in the case of a genetically abnormal fetus. If significant information is lacking, however, the personal biases of the genetic counselor may influence the decision of the family. In this and similar situations, sound information must be obtained before crucial personal or social decisions can be made. The symposium being sponsored by the American Genetic Association is designed to deal with recent developments in human genetics and with resultant social issues. (No refs.) - *B. J. Grylack.*

Johns Hopkins University School of Medicine
Baltimore, Maryland 21205

1630 **DAVIS, BERNARD D.** Prospects for genetic intervention in man. *Science*, 170(3964):1279-1283, 1970.

Recent achievements in the field of molecular genetics have raised controversial questions on the future possibilities of genetic intervention in man. It should be noted that the control of polygenic behavioral traits is much less likely than the cure of monogenic diseases. Moreover, it is pertinent that the most significant human traits, namely intelligence, temperament, and physical structure, are highly polygenic. Thus, while the improvement of cerebral function by polygenic transfer seems extremely unlikely, one cannot exclude so readily the technical possibility of impairing this function by transfer of a monogenic defect. While specific curtailment of genetic research appears to be impossible to justify, one must consider the broader proposal that society may have to limit the rate of scientific progress in general if it hopes to prevent the development of new powers more rapidly than an inadequate institutional framework can handle them. (24 refs.) - *B. J. Grylack.*

Harvard Medical School
Boston, Massachusetts 02115

1631 **UMANSKY, IRVING; WORTH, DOROTHY J.; BENJAMIN, RACHEL; MADOFF, MORTON A.; EDSALL, GEOFFREY; & DIAMOND, LOUIS K.** A state program for prevention of sensitization to Rh factor. *HSMHA Health Reports*, 86(3):212-216, 1971.

Massachusetts General Hospital in Boston initiated a local program to prepare and distribute Rh immune globulin. Voluntary donors, either male or female, had to reside within reasonable proximity to the blood collection center and be sensitized to the Rh factor; female donors had to be beyond the childbearing age or unable to have children. Potential donors had complete physical examinations. An Rh blood profile was performed and Rh antibody level was boosted when necessary. Seventy-nine donors were selected from 434 volunteers. An average donor contributed 550 ml of plasma/month in the program. One liter of plasma yields approximately 51 one ml doses of Rh immune globulin. Plasma fractionated to date has produced 5,418 doses, and that available for fractionation will yield an additional 13,650 doses. Average cost of each 300-microgram dose had been $5.67. Sufficient Rh immune globulin is now available for each woman-at-risk in the state of Massachusetts. (10 refs.) - *J. K. Wyatt.*

Blood Grouping Laboratory
Boston, Massachusetts 02115

1632 **EDWARDS, R. G.; STEPTOE, P. C.; & PURDY, J. M.** Fertilization and cleavage *in vitro* of preovulator human oocytes. *Nature*, 227(5265):1307-1309, 1970.

In vitro fertilization and growth of human oocytes to the 16-cell stage are reported. Spermatozoa were supplied by the husband of each subject from whom ova were obtained. Preovulatory oocytes, in metaphase of the second meiotic division, were most ready for fertilization. Various media were employed, e.g., Bavister's, Whittingham's, Waymouth's, Ham's F10; some modifications were made. Fertilization occurred in all media except Ham's. Fertilized cells were gradually transferred to other media for cleavage. Timing of cleavages in embryos makes it possible to analyze the chromosome complement of the embryo to identify anomalies in early development. (26 refs.) - *E. Kravitz.*

University of Cambridge Physiological Laboratory
Cambridge, England

1633 **HOOK, ERNEST B.; & PETRY, JUDITH J.** Single hit aetiology of human minor congenital malformations unassociated with major congenital malformations. *Nature*, 227(5260):847-848, 1970. (Letter)

An analysis of 200 babies without major malformations indicated that the presence of certain minor anomalies followed a predictable pattern. The probability of clustering was tested by Poisson distribution. A single-hit etiology was predicted. The observed data were in accord with the Poisson-predicted distribution pattern. Data from other studies were also analyzed. (3 refs.) - *E. Kravitz.*

Albany Medical College
New York, New York 12208

1634 **MICHIE, EILEEN A.; & ROBERTSON, JOHN G.** Amniotic and urinary oestriol assays in pregnancies complicated by rhesus immunization. *Journal of Obstetrics and Gynaecology of the British Commonwealth*, 78(1):34-40, 1971.

One hundred and sixty-three samples of amniotic fluid were obtained for estriol determination from 53 Rh negative immunized women with a range of 17 to 38 weeks of gestation in a search for a reliable method of assessing the fetus in pregnancies complicated by rhesus immunization. Urinary estriol was measured in 8 patients before and after the intrauterine transfusion which they required. In most of the pregnancies studied, it was not possible to correlate the estriol concentration in the amniotic fluid with the severity of hemolytic disease in the fetus and infant. A fall in the level of amniotic fluid estriol on serial estimation may be of importance in the diagnosis of a severely affected infant. When the level falls further and is below the normal range, however, metabolic failure in the fetus may be present. Subsequent studies on the estriol concentration in amniotic fluid in cases of Rh immunization are required before a final conclusion can be reached on its efficacy in prognostication. (19 refs.) - *B. J. Grylack.*

University of Edinburgh
Edinburgh, Scotland

1635 **CARTER, C. O.; EVANS, K. A.; *FRASER ROBERTS, J. A.; & BUCK, A. R.** Genetic clinic: A follow-up. *Lancet*, 1:7963, 1971.

A follow-up study of 455 couples interviewed at the genetic clinic of the Hospital for Sick Children in London between 1952 and 1964 sought to determine how well couples understood counseling on risks; what decisions were made on future pregnancies; and the outcome of any further pregnancies. Results indicated that the parents had understood the information they were given, and that most of them made responsible decisions on the basis of the information. Two-thirds of parents who were advised that risk of recurrence was high decided not to plan for any more children; in cases of low risk, only a quarter of the parents were deterred. None of the high risk couples had a planned additional child with a serious long term handicap. Predictions of risk appeared to be accurate on the whole; a proportion of about 1 in 6 of children born to high risk couples were affected, as contrasted to 1 in 50 of the children born to low risk couples. Two cases of erroneous information arising from difficulties of diagnosis are noted. At present genetic counseling has not notably reduced the pool of genetic errors, but its contribution to community health will probably increase steadily. (6 refs.) - *N. Jarvis.*

*Institute of Child Health
London WC1N 1EH, England

1636 Big drama for ill infants. *Medical World News*, 12(12):64, 1971.

The Intermountain Newborn Intensive Care Center in Salt Lake City (Utah) has become a referral center for neonates in need of intensive care. The newborns will be accompanied from their place of birth by trained health personnel and will be transported by ambulance, helicopter, or airplane equipped with the necessary monitoring equipment. Despite the long distances involved in transporting some patients, an improvement in mortality rates for high-risk neonates has been indicated. The Center admitted approximately 200 babies during the past year. However, of the 77,000 births recorded each year in the intermountain region, 700 to 1,500 infants will require intensive care. Plans are being made to expand the capacity of the Center in the near future so that as many as 22 infants can be treated at one time. (No refs.) - *B. J. Grylack.*

1637 **FRASER, F. CLARKE.** Genetic counseling. *Hospital Practice*, 6(1):49-56, 1971.

Genetic counseling has become another facet of medical practice as a result of increasing identification of genetic diseases and better understanding of the mode of inheritance. Of the autosomal recessive, sex-linked recessive, and autosomal dominant modes of inheritance, the most common is autosomal recessive inheritance, found in phenylketonuria, sickle cell anemia, cystic fibrosis, and other diseases. Less common is sex-linked recessive inheritance, found in hemophilia, Duchenne type of muscular dystrophy, and numerous other conditions. Autosomal dominant inheritance is the least common. To provide responsible genetic counseling, the physician must be certain of the diagnosis and must recognize that similar diseases may show different patterns of inheritance. Moreover, he should realize that parental reactions to their children's disease may differ from his own. Both the demand for genetic counseling and the options which are available are increasing as a consequence of changing attitudes towards early interruption of pregnancy and new diagnostic techniques. (No refs.) - *B. J. Grylack.*

McGill University
Montreal, Quebec, Canada

1638 Birth defects and their environmental causes. *Medical World News*, 12(3):47-51, 54-55, 57, 1971.

Recent evidence has documented the damage that specific environmental agents can do to animal fetuses. Notable findings include demonstration of teratogenic and feticidal effects in 2 strains of mice and 1 strain of rats due to the herbicide 2,4,5-trichlorophenoxyacetic (2,4,5-T) acid; adverse genetic effects in mice due to a minor additive in leaded gasoline; and an increase in fetal abnormalities and fatalities in mice and rats due to nitrilotriacetic acid (NTA), a detergent component. There appears to be little doubt that many diseases once regarded as spontaneous are caused by environmental pollutants. The recognition that many human cancers are probably caused by environmental carcinogens and that they are ultimately preventable is growing. It has been proposed that a new interdisciplinary agency be created which is responsive scientifically and legally to a broad range of consumer interests and in which ultimate responsibility for all problems of environmental quality and consumer protection is consolidated. (No refs.) - *B. J. Grylack.*

1639 **HARRIS, MAUREEN.** Mutagenicity of chemicals and drugs. *Science*, 171(3966):51-52, 1971.

Advances in the understanding of genetics which have come from experimental processes have not been achieved without the realization that mutations are occurring in man by processes that are less controlled than those in the laboratory, such as radiation damage and alteration of chromosomes by chemicals and drugs. Geneticists have defined gene mutations and chromosomal breaks or losses as the 2 major categories of chromosomal alteration. Techniques are continually being proposed to determine whether a drug or chemical is mutagenic and what types of mutation it produces. The monitoring of the human population for genetic mutations is an essential part of protecting man from the harmful effects of environmental agents. Hopefully, industry, government, and private foundations will insure long-range guaranteed support for a multigenerational effort in that direction. (No refs.) - *B. J. Grylack.*

No address

1640 Is fetal monitoring worthwhile? *British Medical Journal*, 1(5748):515-516, 1971. (Editorial)

Management of the fetal heart rate and pH of blood are leading aspects of fetal monitoring during labor. The limited use of monitoring in clinical obstetrics is probably due to the anxiety of obstetricians concerning the justification of extra effort and expense rather than to any doubts about the reliability of the technique in detecting fetal asphyxia. The difficulty of interpreting signs of fetal distress is indicated frequently by the tendency among obstetricians to resort to operative delivery. Monitoring can contribute to obstetric management by providing a reliable early warning system for the high-risk fetus and by determining the significance of clinical fetal distress in an otherwise normal pregnancy. Whether fetal monitoring is worthwhile or not depends on the accuracy of monitoring devices and the applicability of the systems to most busy obstetric units. (16 refs.) - *B. J. Grylack.*

1641 **KIRMAN, BRIAN; & ANGELI, EMILY.** Genetic counselling in relation to mental retardation. *Nursing Mirror*, 132(1):21-24, 1971.

Despite a slow but definite increase in information about untoward factors during pregnancy which may interfere with the proper development of the fetus and especially of the brain, the majority of cases of severe MR remain unclassified, and it is still difficult to draw a definite conclusion regarding etiology and to advise the mother appropriately when factors such as rubella are present. The difficult evaluation of perinatal factors in the etiology of MR is a major obstacle to the assessment of genetic risk. In a recent study of a group of SMR children, MR in approximately 20% appeared to be attributable to an external agent, and more than one third of these cases were ascribed to perinatal damage. While ample evidence suggests that environmental factors in the social and cultural group are significant in the causation of mild MR, advice on genetic risks in mild and borderline MR is not always readily accepted. In many cases of high-risk families, children may have been affected by the same genetically determined disease process. Most frequently, the condition responsible for a family pattern of MR cannot be classified. The apparent simplicity of relevant genetic laws may be misleading, and parents may be poorly advised. In the future, advice will be sought more frequently from parents concerned with possible genetic risks, and the professions concerned will have more knowledge of the problem. (No refs.) - *B. J. Grylack.*

Queen Mary's Hospital for Children
Carshalton, England

1642 **DRAZANCIC, ANTE; & STAVLENIC, ANA.** Free fatty acid determinations in normal and abnormal pregnancies. *American Journal of Obstetrics and Gynecology*, 109(4):666-669, 1971.

An elevated free fatty acid (FFA) level in pregnancy, especially at the beginning of a pregnancy, could indicate the prediabetic state and a potentially pathologic pregnancy. FFA determinations were made in four pregnancy groups, including normal pregnancies (62 Ss), pathologic pregnancies with normal glucose tolerance (62 Ss), pathologic pregnancies with prediabetic glucose tolerance (62 Ss), and diabetic pregnancies (18 Ss). The data showed a marked FFA level increase in diabetic pregnancy, slight FFA level rise in normal pregnancy, moderate FFA level increase in pathologic pregnancy with glucose tolerance, and a FFA level in prediabetic pregnancy very close to the diabetic pregnancy level. Of particular significance was the finding that the FFA level in prediabetic pregnancy was already elevated at the beginning of pregnancy. (9 refs.) - *S. Glinsky.*

Zagreb University School of Medicine
Zagreb, Yugoslavia

1643 **SPELLACY, WILLIAM N.; TEOH, ENG SOON; BUHI, WILLIAM C.; BIRK, SHARON A.; & McCREARY, SALLY A.** Value of human chorionic somatomammotropin in managing high-risk pregnancies. *American Journal of Obstetrics and Gynecology*, 109(4):588-598, 1971.

Although human chorionic somatomammotropin (HCS) values in blood correlate with apparent fetal well-being, the clinical usefulness of this test in managing high-risk pregnancies remains open

to speculation. With the use of a solid-phase radioimmunoassay, HCS was measured in 1,378 serum samples from 506 pregnant women. HCS values were similar in the normal, the blood factor Rh-sensitized, and the diabetes mellitus-complicated pregnancies. There was a decrease in the HCS values in proportion to the severity of maternal hypertension. An abnormally low fetal danger zone for HCS was determined, and the high-risk hypertensive pregnancies could be divided into those where serum HCS was above the zone (stillbirth rate zero) and those where the HCS values were in the zone (stillbirth rate 24.1). Significant correlations were found when comparisons were made between serum HCS and urinary estriol excretion or serum heat-stable alkaline phosphatase levels. Hopefully the easy identification of a small group of hypertensive women whose fetuses are at great risk might lead to more intensive obstetric care of this group and an eventual lowering of the perinatal death rate. (32 refs.) - *S. Glinsky.*

University of Miami Medical School
Miami, Florida

1644 **JENKINS, DAVID M.; FARQUHAR, J. B.; & OAKEY, R. E.** Urinary estrogen excretion in prolonged pregnancies. *Obstetrics and Gynecology*, 37(3):442-450, 1971.

Analysis of urinary estrogen excretion may be a valuable means of identifying gravidas for whom the fetus will be at risk if the pregnancy is continued. Data on 184 pregnancies which had passed the expected confinement date were analyzed to study the relationship between placental and fetal condition and estrogen excretion. Estriol and other estrogens were determined by the Kober reaction, and estimates were begun at term or soon after. In uncomplicated preterm pregnancies, estrogen excretion rose progressively with advancing maturity. In complicated pregnancies, the progressive rise in level of estrogen excretion observed during the preterm period did not continue after expected date of confinement but leveled off to a plateau. Heavier babies and larger placentas were associated with higher levels of urinary estrogen excretion. Low and falling levels of estrogen seemed related to reduced resistance to the hypoxic stress of labor. When serious placental insufficiency is suspected, continuous estimates of urinary estrogen levels should be made. The results of each 24-hour collection should be available within 6 to 8 hours. (21 refs.) - *J. K. Wyatt.*

University of Leeds
Leeds, LS 2, 9 NG, England

1645 **KIRMAN, BRIAN.** The aetiology of mental subnormality. *Modern Perspectives in Child Psychiatry*, 1:496-522, 1971.

Many of the causes of MR are still unknown, but many genetically determined conditions and social and environmental factors have been defined. In the absence of precise etiology, all factors which improve the health and cultural level of the community, especially mother and child, will probably reduce the incidence of MR. Remedial and palliative measures for established defects will also reduce the burden on the family and community. True prevention depends upon further research into etiology; extensive investigative programs are necessary. (89 refs.) - *E. L. Rowan.*

Fountain and Carshalton Hospital Group
Surrey, England

1646 **ROSZKOWSKI, IRENAEUSZ; IWANSKA, JANINA; MYSZKOWSKI, LEOPOLD; BRZESKI, JERZY; & SCHMALHOFER, ALICJA.** Blood serum amino acids of non-pregnant women who have given birth to children with gross developmental malformations. *American Journal of Obstetrics and Gynecology*, 108(8):1213-1216, 1970.

The concentrations of amino acids in the blood sera of non-pregnant women and men whose children had congenital malformations differed from amino acid concentrations in parents with normal children. The Ss were 23 women and 17 men with children afflicted with gross developmental malformations and a control group of 22 mothers and 22 fathers of at least 2 healthy children. Quantitative chromatography of the amino acids in blood sera was done by the starch column-paper method. Of the 8 amino acids in the women's bloood sera and 10 amino acids in the men's blood sera showing differences, the majority of the cases showed marked increases in

threonine and glutamic acid. Such amino acid changes seem to predispose to birth of a malformed child. (10 refs.) - *S. Glinsky.*

Medical Academy
Warsaw, Poland

1647 **WALKER, JOHN.** Mortality in low birthweight infants. *Developmental Medicine and Child Neurology,* 12(5):661-663, 1970.

Analysis of social and economic factors in maternal history is an accurate indicator in identifying high risk mothers who may bear low birthweight infants. Greater population mobility and the prospect of lower future birth rates necessitate less reliance on a general social class factor and past obstetrical performance. A study of 12,000 deliveries in northern Finland utilized 47 biological and social variables, none dependent on clinical examination and all potentially available by questionnaire. These variables were then related to obstetric performance measured by perinatal mortality, birthweight, gestational age, and intrauterine growth. The clinical findings reemphasized the importance of low birthweight and immaturity as the major factors in perinatal mortality. The 37 social and economic factors of social class varied considerably by weighting, but all analyses proved effective in isolating the under 2,500g group, which is repeatedly at highest risk. Such effective predictive information can be easily obtained by a simple questionnaire. Computer correlation of this as well as of additional perinatal, neonatal, developmental, and social information can add precision to an aspect of preventive care which has used crude methods of measurement far too long. (2 refs.) - *S. Glinsky.*

University of Newcastle upon Tyne
Medical School
Newcastle upon Tyne NE1 7RU, England

1648 **SWINYARD, CHESTER A.** Genetic counselling in rehabilitation medicine. *International Rehabilitation Review,* 22(1):3-7, 1971.

Sound genetic counseling rests upon a basic understanding of human inheritance mechanisms. All genetic information is transferred to the fertilized egg cell or zygote by mitosis of DNA molecules known as chromosomes. Every normal individual possesses 46 chromosomes, which comprise his karyotype; hereditary defects may be due either to a defective gene or to the loss or addition of a complete chromosome. A third type of defect may be attributed to several gene defects and is called polygenic or multifactorial inheritance. Gene defects, in addition to being hereditary, may also be caused by environmental factors or internal pressures and are then classified as mutations. Genetic counseling depends entirely upon an accurate medical and genetic diagnosis. Parental questions regarding the advisability of having more children can be answered relatively easily if the condition is one transmitted in one of the Mendelian ratios, but the question is more complex if the disease is one of multifactorial inheritance. Much can be done with a hereditary disease to preserve life, reduce morbidity, and enable patients to achieve independence. Amniocentesis makes possible the detection of disease before birth of the child. Genetic counseling must be done on an on-going, periodic basis, since the genetic implications in an individual case become clearer with increasing age. (10 refs.) - *S. Glinsky.*

New York University Medical Center
New York, New York

1649 **HENNEMAN, CAROL E.; ANDERSON, GAIL V.; TEJAVEJ, ANANT; GROSS, HOWARD A.; & HEIMAN, MELVIN L.** Fetal maturation and amniotic fluid. *American Journal of Obstetrics and Gynecology,* 108(2):302-307, 1970.

Amniotic fluid creatinine, bilirubin, and cells as well as serum creatinine were measured to corroborate their reported correlations with fetal maturation. Results show that an amniotic fluid creatinine value of 2 mg per 100 ml or more usually indicates a gestational age of at least 37 weeks. Absence of a bilirubin peak at 450 mμ, the presence of 20% of cells which stain orange with Nile blue sulfate dye, or the presence of a ratio between amniotic fluid creatinine and serum creatinine of 3 or more usually indicates a gestational age of 36 or more weeks. A high percentage of these tests are negative beyond 36 to 37 weeks. At present these measurements have limited clinical value because no single determination or combination of determinations provides a

reliable index of fetal maturation. On the other hand, amniotic fluid analysis is a useful supplement to other criteria of fetal maturity in those cases where an accurate estimate of gestational age is of critical importance. (9 refs.) - *J. C. Moody.*

University of Southern California Medical Center
Los Angeles County, California

MEDICAL ASPECTS — Etiologic Groupings

Infections, intoxication, and hemolytic disorders

1650 WALKER, W. Phenobarbitone in rhesus haemolytic disease. *Lancet*, 2(7682):1089-1090, 1970. (Letter)

The role of phenobarbitone in the management of Rh-isoimmunization may be in reducing the need for repeated transfusions rather than as the first step in treatment. Only 30% of Coombs-positive infants will develop jaundice severe enough to require exchange transfusion; the role of phenobarbitone in lowering this percentage is questionable. (1 ref.) - *E. L. Rowan.*

Royal Victoria Infirmary
Newcastle upon Tyne, England

1651 SCHOENBAUM, STEPHEN C. Problems of rubella control. *New England Journal of Medicine*, 283(15):819, 1970. (Letter)

The criterion of vaccine effectiveness must be immediately added to those of safety and potency. Scientific evidence that rubella vaccination is effective in preventing congenital rubella malformations must be developed now. To continue an unproved mass vaccination program is experimentation in its least beneficial and most hazardous form. (4 refs.) - *G. S. Bell.*

Beth Israel Hospital and
Children's Hospital Center
Boston, Massachusetts

1652 KEMBALL, M. L.; McIVER, C.; MILNER, R.D.G.; NOURSE, C. H.; SCHIFF, D.; & TIERNAN, J. R. Neonatal hypoglycaemia in infants of diabetic mothers given sulphonylurea drugs in pregnancy. *Archives of Disease in Childhood*, 45(243):696-701, 1970.

Sulphonylurea drugs should not be utilized to manage diabetes mellitus of a mother during pregnancy. Of 4 infants whose diabetic mothers were given these drugs (chlorpropamide for 3 and acetohexamide for 1) during pregnancy, all developed severe hyperinsulinemia, which caused severe hypoglycemia during the neonatal period. Correction of this condition required vigorous and prolonged treatment, including exchange transfusions for 2 to lower the concentration of the drug. In the mother receiving acetohexamide, treatment was continued throughout the pregnancy; however, the 3 receiving chlorpropamide were treated with this drug from 34 weeks to delivery (at 36 weeks of gestation), from 31 weeks to delivery (at 38 weeks), and from 22 weeks to delivery (at 36 weeks), respectively. Examination of glucose, insulin, and drug levels in the 4 infants showed that the drugs had crossed the placental barrier, and their fetal plasma concentrations were in the therapeutic range for diabetic adults. Besides use of exchange transfusions in 2 of the infants, management of the hypoglycemia included intravenous glucose administration, addition of fructose to the diet of 1 S, and exclusion of leucine from the diet of another. (32 refs.) - *M. S. Fish.*

Hammersmith Hospital
Du Cane Road, London W.12, England

1653 AMATAYAKUL, O.; CUMMING, G. R.; & *HAWORTH, J. C. Association of hypoglycemia with cardiac enlargement and heart failure in newborn infants. *Archives of Disease in Childhood*, 45(243):717-720, 1970.

A review of chest X-rays of infants with neonatal hypoglycemia has indicated that this condition is frequently associated with cardiac enlargement and heart failure. Of 43 newborn infants with

hypoglycemia (not including infants of diabetic mothers), 16 had undergone chest radiography; of these, 12 who were born at term had X-rays which showed cardiac enlargement. Four of the 12 (3 males, 1 female) had associated cardiac failure and abnormal electrocardiograms. One died at 7 days of cerebral hemorrhage; 2 were MR at 22 months and 3 years, respectively; and 1 was normal at 1 year. Of the remaining 8 (5 males, 3 females) without cardiac failure, all recovered; however, 1 was MR at 1 year, and 1 had recurrent hypoglycemia. All infants were treated with intravenous infusions of glucose, and corticosteroids were given when blood glucose levels remained low. Response to glucose was much better in the group without cardiac failure. Of 8 infants examined on follow-up, all had normal hearts. While the results do not imply a cause and effect relationship between hypoglycemia and cardiac enlargement and failure, they do suggest that when 1 of these abnormalities occurs in the newborn, the infant should be examined to determine if the other is present also. (9 refs.) - *M. S. Fish.*

*The Children's Hospital
Winnipeg 3, Manitoba, Canada

1654 **McENERY, G.; & STERN, H.** Cytomegalovirus infection in early infancy. Five atypical cases. *Archives of Disease in Childhood,* 45(243):669-673, 1970.

A reported occurrence of 5 cases of cytomegalovirus infection, with none showing the classical symptoms, in a neonatal ward over a 2-year period indicates the possibility that this type of infection is more widespread than previously believed and that clinical features may often be milder than those usually observed in these cases. Of the 5 cases, 2 were associated with the rhesus incompatibility, and 2 were initially diagnosed as ABO incompatibility. These 4 cases were given exchange transfusions; 1 of the infants with rhesus isoimmunization died. A fifth infant, who also died, had skeletal and corneal anomalies but no symptoms of cytomegalovirus infection, which was found only at autopsy and only in the salivary glands. Evidence of the virus infection was found in the urine of the 3 surviving infants. In these latter cases positive viral tests in the mother and negative tests for the donor blood indicated that the infection was congenital. The infant with the congenital anomalies probably acquired the infection about the time of birth, and no conclusions could be drawn about the source of infection in the other infant who died. Although 2 of the survivors had small heads, mental development was normal at 7 and 16 months, respectively. (16 refs.) - *M. S. Fish.*

Hammersmith Hospital
London W.12, England

1655 **WALLACE, SHEILA J.; & ZEALLEY, HELEN.** Neurological, electroencephalographic, and virological findings in febrile children. *Archives of Disease in Childhood,* 45(243):611-623, 1970.

Studies of febrile children have demonstrated a significantly more frequent occurrence of permanent neurological damage and of electroencephalographic (EEG) abnormalities among those with viral disease compared to those without such infections. Severe fits were also more common among the virus-infected group. Of 78 febrile children, ages from 1 month to less than 7 years on admission for study, 28 with fits had positive viral findings; 25 with fits had negative viral findings; 18 control Ss with no history of convulsions had positive and 7 control children had negative viral findings. Studies of the birth, family, and developmental histories, evidence of earlier neurological damage, and clinical, neurological, and viral investigations disclosed that 53% of the children with fits had viral illnesses, including those already known to cause neurological damage and several not previously associated with neurological complications: adenovirus 7, respiratory syncytial virus, parainfluenza 2, and *C. burnetti* (Q fever). Viral infections appeared to be more important than fits in determining speed of the dominant EEG rhythm and its return to normal. The results indicate that encephalitis may be a more frequent concomitant of febrile convulsions than is now commonly believed and that apparently innocuous childhood illnesses can cause cerebral dysfunction which, later, may be associated with epilepsy and with motor and behavior disturbances. (34 refs.) - *M. S. Fish.*

Cardiff Royal Infirmary
Cardiff, CF2 1SZ, Wales

1656 **SILBERBERG, DONALD H.; JOHNSON, LOIS; SCHUTTA, HENRY; & RITTER,**

LINDA. Effects of photodegradation products of bilirubin on myelinating cerebellum cultures. *Journal of Pediatrics*, 77(4):613-618, 1970.

When cultures of myelinating cerebellum were exposed to unirradiated media containing bilirubin and human serum albumin, characteristic and severe damage was produced. Exposure to identical media in which the bilirubin was partially degraded by light, and remaining diazotizable bilirubin was below 14 mg%, did not result in damage to the culture. The bilirubin level was 30%/100 ml; final albumin concentration in the medium was 2.0 gm/100 ml. A relatively constant rate of decrease in the level of bilirubin was found when cultures of myelinating cerebellum were exposed to irradiated media containing bilirubin and human serum albumin. Fifty-one cultures were exposed to unirradiated medium containing bilirubin, 103 cultures were exposed to irradiated medium containing bilirubin, and 61 cultures were exposed to irradiated medium which did not contain bilirubin. Irradiation was effected by exposure to two 15-watt blue fluorescent bulbs at a distance of 4 inches for periods of 3, 4, 6, or 24 hours. No cytotoxicity was observed in the photodegradation products, indicating that phototherapy is safe to use for the prevention of kernicterus. (11 refs.) - *J. K. Wyatt.*

University of Pennsylvania School of Medicine
Philadelphia, Pennsylvania

1657 ODIEVRE, M.; GAUTIER, M.; NOCTON, F.; & ALAGILLE, D. Cholostase intrahepatique par obstruction des voies biliaires interlobulaires au cours d'une maladie generalisee des inclusions cytomegaliques chez un nouveau-ne (Intrahepatic cholestasis through obstruction of the interlobular biliary passages in the course of a generalized cytomegalic inclusion disease in a neonate). *Archives Francaises de Pediatrie*, 27(2):211-216, 1970.

Congenital cytomegalic inclusion disease complicated by intrahepatic cholestasis was manifested in a neonate and was confirmed rapidly by the discovery of inclusion cells in the urinary residuum and by the presence of the cytomegalic virus elevated in the urine and liver. No intrahepatic biliary atresia was noted. The most unique characteristic was the cholestatic evolution verified in the third week of life. The cholestasis was marked by discoloration of the stools, while jaundice was observed with hyperbilirubinemia and with a progressive cholesterololipidic retention. The occurrence of cholestasis has already been described in the course of congenital rubeola. However, histologic examination of the liver, which evidenced luminal obstruction by cytomegalic epithelial cells of intraportal biliary ducts and which displayed all the characteristics of cytomegalia, manifested a certain originality of the biliary obstruction mechanism. (4 refs.) - *B. J. Grylack.*

Unite de Recherche d'Hepatologie Infantile
Hopital Parrot
94 - Bicetre, France

1658 McMULLIN, G. P.; HAYES, M. F.; & ARORA, S. C. Phenobarbitone in rhesus haemolytic disease. A controlled trial. *Lancet*, 2(7680):949-952, 1970.

Treatment and control groups consisting of 30 infants each were used in a study of the role of phenobarbitone in the management of infants with rhesus hemolytic disease. Phenobarbitone was administered every 8 hours in a dose of 8 mg to Ss over 6 lb 8 oz and in a dose of 1 mg/lb to Ss under 6 lb 8 oz within the treatment group after the result of the cord-blood Coombs test was known. Exchange transfusion was performed for a member of either group if the cord-blood serum bilirubin was above 4 mg per 100 ml, if cord-blood hemoglobin was less than 13 g per 100 ml, or if capillary blood serum bilirubin was above 20 mg per 100 ml, assuming a positive Coombs test in all cases. Thirty control infants required 40 exchange transfusions and 30 treated infants required 21 exchange transfusions, amounting to a 2% significant difference level. If the immediate exchange transfusions at birth are excluded from consideration, the difference is highly significant. The relationship between the use of phenobarbitone in the neonatal stage and clinical events later in life is a matter of speculation at this time. (24 refs.) - *B. J. Grylack.*

Children's Hospital
Sheffield S10 2TH, England

1659 **GULBIS, E.; WEBER, A.; DECHAMPS, L.; *DENYS, P.; SOKAL, G.; LOHR, G.; RUDIGER, H.; BLUME, K.; PIRET, L.; & DUNJIC, A.** Contribution a l'etude de l'anemie hemolytique congenitale avec deficit en pyruvate-kinase (Contribution to the study of congenital hemolytic anemia with a pyruvate-kinase deficiency). *Archives Francaises de Pediatrie*, 27(1):31-49, 1970.

Severe congenital non-spherocytic hemolytic anemia was followed in a male child until the age of 3 years 6 months. Numerous transfusions were necessary despite an active erythropoiesis. Aggravations coincided regularly with rhinopharyngeal and bronchial infections in the S in the absence of apparent alteration of the immunological system. Erythrocytic pyruvate-kinase activity was a third of the normal value, adenosine triphosphate (ATP) content was normal, and the remaining pyruvate-kinase was qualitatively abnormal. Genetic study of the family indicated that only 1 out of 7 siblings had a normal erythrocytic pyruvate-kinase. The parents, 2 brothers, and 1 sister of the S were found to be heterozygous carriers of the defect. The finding that the decline of the disease is masked partially by the abundance of reticulocytes explains the appearance of the defect in homozygous and not in heterozygous Ss, although the difference in pyruvate-kinase activity between the 2 groups does not appear to be significant. (34 refs.) - *B. J. Grylack.*

*Universite de Louvain
Louvain, Belgium

1660 **TOIVANEN, P.; & HIRVONEN, T.** Sex ratio of newborns: Preponderance of males in toxemia of pregnancy. *Science*, 170(3954):187-188, 1970.

On the basis of data collected from obstetrical records of 1,061 mothers with toxemia and 8,257 controls, the hypothesis of incompatibility of fetus and mother as a cause of toxemia of pregnancy was examined. Of the mothers with toxemia, there was a 1.24 ratio of those delivering males to those delivering females, a ratio which increased to 1.71 when urinary output of protein exceeded 3 g per 24 hours. It appears that the increased ratio of males to females born to toxemic mothers is an expression of histoincompatibility between mother and fetus. Like other paternal antigens in all placentas, the potential Y-chromosome dependent antigen(s) must be present in the placenta of the male fetus. Histoincompatibility antigens may potentiate the immunogenicity of other placental antigens known to be shared by the kidney, and this immunization may lead to renal lesions and other characteristic features of toxemia. (26 refs.) - *B. J. Grylack.*

University of Minnesota
Minneapolis, Minnesota 55455

1661 Two-pronged attack on pica. *Medical World News*, 11(33):32, 1970.

While the Congress contemplates legislation against lead-based paints, individual cities are attempting to attack the problem of pica on a local level. In 1970 New York City established the Bureau of Lead Poisoning Control within its own health department. As a result, there has been a fourfold increase in the detection of children with at least 0.06mg% of lead in their blood. Prevention of lead poisoning is being emphasized, and a housing repair requirement is being enacted together with a case finding operation. An educational program has been initiated through public health personnel activity and a media advertising campaign. In the first 6½ months of the program, the Bureau has tested more than 30,000 children. (No refs.) - *B. J. Grylack.*

1662 **MATHERS, N. P.; JAMES, G. B.; & WALKER, J.** The oxygen affinity of the blood of infants treated by intrauterine transfusion. *Journal of Obstetrics and Gynaecology of the British Commonwealth*, 77(7):648-653, 1970.

Six mothers receiving intrauterine transfusions for rhesus hemolytic disease and 7 normal patients were used to study the oxygen-carrying properties of adult blood within the fetal environment and to establish whether that environment alters the oxygen affinity of the adult red cell and thus maintains the normal relationship between the maternal and fetal bloods. On the basis of umbilical vein and maternal bloods and the measurement of total hemoglobin, fetal

hemoglobin, percentage of fetal red blood cells, and oxygen dissociation curves and oxygen affinity, it was found that the adult red cells retained their adult-type oxygen affinity when transfused into the fetal environment and that the fetuses were able to survive and, apparently, to grow normally for up to 6 weeks. Of the various factors involved, it appears that only the oxygen tension distinguishing between maternal and fetal blood is essential for survival. (22 refs.) - *B. J. Grylack.*

University of Dundee
Dundee, Scotland

1663 **PORTO, SERGIO O.; PILDES, ROSITA S.; & GOODMAN, HAROLD.** Relationship of phototherapy and skin color. *Journal of Pediatrics*, 77(6):1099, 1970. (Letter)

"Daylight" and "Vitalite" fluorescent lamps have much less energy than blue fluorescent lamps. Blue lamps should be used at least 75% of the time for adequate phototherapy in Negro infants, and contradictory reports of success with this method of therapy should be reevaluated in terms of the sources of light. (4 refs.) - *E. L. Rowan.*

Grant Hospital
Maywood and Chicago, Illinois

1664 **ENTE, GERALD.** Relationship of phototherapy and skin color. *Journal of Pediatrics*, 77(6):1098, 1970. (Letter)

Increasing the duration of phototherapy reduced the peak serum bilirubin concentration in Caucasian babies (N=41) but not in Negro babies (N=55). One Negro infant required exchange transfusion despite this treatment. "Cool white" fluorescent lighting was used in this study and may account for the differences from previous reports; however, phototherapy should still be used with caution in jaundiced Negro infants. (1 ref.) - *E. L. Rowan.*

Meadowbrook Hospital
East Meadow, New York 11554

1665 **STAUB, HENRY P.** Rubella as a possible cause of diastrophic dwarfism. *Journal of Pediatrics*, 77(6):1096-1097, 1970. (Letter)

Diastrophic dwarfism was noted in an infant with a positive rubella culture at birth. A positive titer at age 8 months and a clinical history of maternal rash and positive contact during the third month of pregnancy lend further support to a congenital rubella infection. Despite the marked foreshortening of extremities and complete cleft palate the child showed normal developmental milestones. Diastrophic dwarfism is considered an autosomal recessive disease and establishment of an other than coincidental relationship with rubella will require further case reports. (2 refs.) - *E. L. Rowan.*

E. J. Meyer Memorial Hospital
Buffalo, New York 14215

1666 **PATEL, DAKSHA A.; PILDES, ROSITA S.; & BEHRMAN, RICHARD E.** Failure of phototherapy to reduce serum bilirubin in newborn infants. *Journal of Pediatrics*, 77(6):1048-1051, 1970.

Eight newborn infants with hyperbilirubinemia failed to show a response (a drop of 3 to 4 mg/100 ml after 24 hours of therapy) to phototherapy. Five of the cases had OA incompatibility, 1 Rh incompatibility, and 2 idiopathic hyperbilirubinemia and prematurity. Phototherapy may not always reduce neonatal jaundice, and such infants should be monitored carefully in order to initiate exchange transfusions when indicated. (9 refs.) - *E. L. Rowan.*

700 S. Wood St.
Chicago, Illinois 60612

1667 **ACKERMAN, BRUCE D.** Phenobarbital for neonatal jaundice. *Journal of Pediatrics*, 77(5):916-917, 1970. (Letter)

Phenobarbital cannot be recommended as a treatment for neonatal jaundice because of its respiratory complications. Previous studies have shown a paradoxical response to phenobarbital anesthesia in newborn dogs. They hypoventilated in

response to metabolic acidosis rather than hyperventilating as did adult dogs. This response added respiratory acidosis to the already critical clinical picture. (4 refs.) - *E. L. Rowan.*

University of California
Irvine, California

1668 YEUNG, C. Y. Hypoglycemia in neonatal sepsis. *Journal of Pediatrics,* 77(5):812-817, 1970.

Seventeen of 56 infants admitted to hospital with obvious sepsis were also found to have hypoglycemia. All responded to glucose infusion. Three other infants whose blood sugar was monitored showed hypoglycemia before death and pneumonia at autopsy. Gram-negative bacteria were incriminated in infections associated with hypoglycemia, and gram-positive bacteria in infections with normoglycemia. The mechanism of hypoglycemia in sepsis is unknown. (21 refs.) - *E. L. Rowan.*

Queen Mary Hospital
Hong Kong

1669 AMMANN, A. J.; ROTH, J.; & HONG, R. Recurrent sinopulmonary infections, mental retardation, and combined IgA and IgE deficiency. *Journal of Pediatrics,* 77(5):802-804, 1970.

A 5½-year-old girl with MR and recurrent sinopulmonary infections was noted to have deficiencies of both IgA and IgE. Humoral antibody response and cellular immunity were normal. There was no evidence of ataxia-telangiectasia. High titers of anti-IgA antibodies were noted. (7 refs.) - *E. L. Rowan.*

University of Wisconsin Medical Center
Madison, Wisconsin

1670 HURTER, PETER; SCHROTER, WERNER; SCHEDEL, INGOLF; & GERCKEN, GUNTHER. Phospholipids of red cells and blood plasma in adults, newborn infants, and patients with Rh erythroblastosis. *Pediatrics,* 46(2):259-266, 1970.

Phospholipid concentrations of red blood cells and plasma were studied in blood from 10 healthy adults, 6 newborn infants with erythroblastosis due to Rh incompatibility, and the cords of 10 full-term neonates. Normal fetal red blood cells and plasma showed a reduction of total phospholipids and decreases of individual plasma phospholipids; red blood cell phosphatidylcholine, phosphatidylethanolamine, and phosphatidylserine were reduced; sphingomyelin was unchanged. These decreases were less marked in the red blood cells of erythroblastotic infants, but plasma phospholipid patterns were similar. (26 refs.) - *E. Kravitz.*

University of Hamburg
Hamburg, Germany

1671 Rubella vaccine reminder: Up to 15% of children shed. *Medical World News,* 11(39):6, 1970.

The government is adhering to its policy of recommending rubella vaccine for children between 1 year and puberty, with priority for kindergarten and primary schoolchildren, despite new observations regarding the increasing numbers of children who are shedding virus, significance of reinfection, and untoward reactions. Revised recommendations accompanying the policy note that up to 15% of children have had rubella-like symptoms as a result of viral shedding, which can occur for up to 4 weeks following vaccination, but at apparently low hazard to pregnant women. Reinfection tends to occur in the case of vaccinees with low antibody titers but there is no evidence that reinfected vaccinees can transmit virus to susceptible contacts. (No refs.) - *B. J. Grylack.*

1672 CROSBY, WARREN M.; BROBMANN, GUENTHER F.; & CHANG, ALVIN C. K. Intrauterine transfusion and fetal death. *American Journal of Obstetrics and Gynecology,* 108(1):135-138, 1970.

Experiments with 3 fetal baboons (138, 147, and 167 days gestation) confirm the hypothesis that fetal intraperitoneal transfusion may elevate intraperitoneal pressure above that in the umbilical vein, producing fetal death by anoxia. Dye containing fluid was infused intraperitoneally

into the fetal baboons and umbilical vein pressure was measured directly at 14.5 mm Hg. In 2 fetuses, infusions of 23 and 25 ml fluid were needed to elevate the intraperitoneal pressure above 5 mm Hg, and umbilical flow ceased at this pressure. The third fetus required 65 ml to elevate intraperitoneal pressure to 13 mm, and at that pressure umbilical flow ceased. The infusion was continued in the third fetus, and death resulted. Fetal intraperitoneal pressure should be carefully monitored during human intrauterine fetal transfusion. (3 refs.) - *J. K. Wyatt.*

University of Oklahoma Medical Center
Oklahoma City, Oklahoma

1673 **GUSDON, JOHN P.; LEAKE, NORMAN H.; PRICHARD, ROBERT W.; & RHYNE, A. LEONARD.** Amniotic fluid antibody titers and other prognostic parameters in erythroblastosis. *American Journal of Obstetrics and Gynecology*, 108(1):85-90, 1970.

Amniotic fluids and maternal sera from 54 patients within 2 weeks of expected delivery were analyzed to determine weeks of gestation, Δ OD_{450} % T 520/% T 490 ratio, amniotic fluid indirect Coombs titer, maternal serum indirect Coombs titer, ratio of maternal serum indirect Coombs titer and amniotic fluid titer, and the bilirubin/protein ratio. A comparison was made of each factor and all possible combination factors and their ability to predict fetal outcome. All amniotic fluids were tested by immunodiffusion for IgM. The most effective prognostic methods in order of significance were % T 520/% T 490 ratio, Δ OD_{450}, and the amniotic fluid antibody titer. The prognostic effectiveness of any single factor improved when factors were combined in a formula derived from multiple stepwise regression analyses of all possible combinations of factors under consideration. (36 refs.) - *J. K. Wyatt.*

Wake Forest University
Wake Forest, North Carolina

1674 **HAIRE, MARGARET; & HADDEN, DIANA S. M.** Immunoglobulin responses in rubella and its complications. *British Medical Journal*, 3(5715):130-132, 1970.

Assessment of normal responses of rubella-specific IgG and IgM antibody in 8 patients by immunofluorescence revealed a prolonged rubella-specific response in 3 cases with complications of rubella infection. One patient had carpal-tunnel compression, and there were 2 cases of thrombocytopenic purpura. These findings suggest that persistence of infection or alteration of the immunological response may be factors in pathogenesis. A differentiation, similar to that involving immunocompetent cells in the embryo when infection occurs early in pregnancy, may occur in some cases in the mature individual. (30 refs.) - *J. K. Wyatt.*

Queen's University of Belfast
BT12 6BN, Northern Ireland

1675 **REID, WINIFRED M.** Congenital rubella—one approach to prevention. *Canadian Nurse*, 67(1):38-40, 1971.

At Burnaby General Hospital in Canada, following the birth of a boy to a mother who contracted rubella in early pregnancy and the discovery that the child was excreting rubella virus, all pediatric and obstetric nursing staff were given the hemagglutination inhibition test (HI) to determine immune status to rubella. Results showed 18% of those tested to be essentially negative (a titer of less than 1:8). Rubella may be expected to reach epidemic proportions every 6 to 9 years. Since it is highly contagious, pediatric units don't admit cases unless absolutely necessary because of complications. Young married nurses must be protected. Research has shown that in rubella contracted 3 to 4 weeks after the last menstrual cycle there is a 60% chance of anomalies in the infant; in rubella contracted 13-16 weeks after the cycle there is a 7% chance. Since the baby can excrete the virus for a year or 2, there is a hazard for hospital personnel. Gamma globulin should be restricted to susceptible mothers exposed to rubella in whom there is yet no clinical evidence of the infection. Therapeutic abortions are advisable in some cases. A live attenuated virus vaccine is now being used to control impending epidemics, but, since the virus can cross the placental barrier, the vaccine should be used in sexually active women of child-bearing age only if pregnancy can be excluded. (8 refs.) - *B. Berman.*

Burnaby General Hospital
Burnaby, British Columbia, Canada

1676 Laboratory diagnosis of rubella. *British Medical Journal*, 3(5717):237-238, 1970. (Editorial)

Inasmuch as laboratory assessment of more than 200 patients exposed to or developing rubella-like illness during pregnancy has shown that clinical diagnosis was accurate in less than 50% of cases, the need for virological testing of these patients is urgent. Laboratory diagnosis of rubella, achieved primarily through serological tests, should be performed as early as possible. The hemagglutination-inhibition test is preferred for its reliability and sensitivity, but it requires serum samples taken within 48 hours of the onset of symptoms or within 2 weeks of exposure to detect diagnostic rises of antibody titer in convalescent sera. Investigation of patients presenting later is accomplished with the complement-fixation test and the determination of rubella-specific macroglobulin (IgM) antibody. It is often difficult to interpret the results of tests on sera from patients presenting late after infection, however. The indirect fluorescent antibody test and the neutralization test are less widely used. If the routine vaccination in adolescence against rubella now being offered in Great Britain is successful, the need for virological tests will decline. (9 refs.) - *B. J. Grylack.*

1677 Gammaglobulin in rubella. *British Medical Journal*, 3(5718):332, 1970.

The thesis that gammaglobulin administered as a prophylactic against rubella in early pregnancy diminishes the number of fetal abnormalities is difficult to prove. A follow-up study approximately 2 years following administration of gammaglobulin to women exposed to rubella in pregnancy showed that of those given gammaglobulin whose contact with the disease arose within the family, 1.95% developed clinical signs of rubella, as compared with 0.48% whose contact with the disease was outside the family. When the women who had been in contact with rubella in pregnancy were compared with those who received gammaglobulin after contact with the disease, it was found that therapy had probably reduced stillbirths from 1.7 to 1.3% and congenital defects from 2.9 to 2.4%. Those with definite clinical rubella in the first 12 weeks of pregnancy produced approximately a quarter of their children with some abnormality. It seems probable, therefore, that gammaglobulin does reduce the incidence of congenital defects in mothers exposed to rubella in early pregnancy, but this therapy cannot be relied on completely. At the present time, virological testing can help in determining whether a contact of rubella has recently had an infection by the virus. If infection can be confirmed, the pregnancy should be terminated. (3 refs.) - *B. J. Grylack.*

1678 **THALER, M. MICHAEL.** Substrate-induced conjugation of bilirubin in genetically deficient newborn rats. *Science*, 170(3957):555-556, 1970.

Postnatal bilirubin uridine diphosphate-glucuronyltransferase (BGT) activity was studied in heterozygous offspring of a mutant Wistar strain of rats with an autosomal recessive defect in bilirubin conjugation in order to examine the role of substrate induction in the perinatal development of the bilirubin conjugating system. The results indicated that embryonic differentiation causes the elaboration of sufficient BGT activity to prevent jaundice in normal but not in partially deficient newborns. Heterozygotes exposed to large amounts of bilirubin throughout gestation conjugate the pigment twice as efficiently as those in similar but unexposed litters. The presence of bilirubin is associated with accelerated induction of BGT before and after birth. Transient hyperbilirubinemia in premature infants appears to reflect a perinatal adaptive process necessary for eventual survival. (11 refs.) - *B. J. Grylack.*

University of California
San Francisco, California 94122

1679 **ZLOTNICK, A.; & FRIED, K.** Sea-blue histiocyte syndrome. *Lancet*, 2(7676):776, 1970. (Letter)

A Jewish family from Iran was seen with the sea-blue histiocyte syndrome involving both parents and 2 of their children. Typical sea-blue histiocytes and foam-cells were found in the bone marrow of the children and father and in a liver biopsy specimen from the mother, who refused a bone marrow aspiration. This disease appears to be transmitted by an autosomal recessive gene, with typical cells always present in

homozygotes but only occasionally in heterozygotes. (5 refs.) - *B. J. Grylack.*

Hadassah University Hospital
Jerusalem, Israel

1680 **SPIRER, Z.; & BOGAIR, N.** Blood-sugars in infants with diarrhoea. *Lancet*, 2(7686):1311-1312, 1970. (Letter)

During the period from 1967 to 1969, 70 infants were seen for treatment of acute dehydration due to diarrhea and vomiting. Particular attention was paid to the blood sugar levels in relation to this clinical condition. In a patient population with a mean CA of 2.5 months, 17 out of 23 patients whose carbon dioxide (CO_2) content was less than 11 meg/litre were found to be hyperglycemic. Of the 22 hypernatremic infants, only 5 were hyperglycemic. It appears that acidosis is a more constant factor than hypernatremia in the causation of hyperglycemia. It seems probable that both acidosis and hypernatremia produce hyperglycemia by interfering with cellular glucose absorption and utilization, perhaps by inhibiting phosphorylation processes. Hypoglycemia as well as hypocalcemia and hyponatremia should be looked for in cases of post-acidotic convulsions. (8 refs.) - *B. J. Grylack.*

Hadassah Hospital
Tel-Aviv, Israel

1681 **LEVI, JONATHAN; GATMAITAN, ZENAIDA; & ARIAS, IRWIN M.** Deficiency of hepatic organic anion-binding protein, impaired organic anion uptake by liver and "physiologic" jaundice in newborn monkeys. *New England Journal of Medicine*, 283(21):1136-1139, 1970.

This study in monkeys indicated the possible existence of an etiologic relationship between a deficiency of protein Y, a major acceptor protein, and physiologic jaundice in newborn animals. It required 10 days for the neonates to reach normal values for bromsulfalein, plasma clearance, serum bilirubin, and hepatic intracellular Y. Protein Y is described as a cytoplasmic organic anion-binding protein. There seemed to be no relationship between this type of jaundice and protein Z. (28 refs.) - *E. Kravitz.*

Albert Einstein College of Medicine
Bronx, New York 10461

1682 **TIMMONS, G. D.; & JOHNSON, KENNETH P.** Aqueductal stenosis and hydrocephalus after mumps encephalitis. *New England Journal of Medicine*, 283(27):1505-1507, 1970.

The case of a 5-year-old boy who developed aqueductal stenosis 27 months after an episode of mumps encephalitis is described. The mumps diagnosis was confirmed by a rise in complement fixation titer from zero during the acute phase to 1:256 (soluble antigen) and 1:512 (viral antigen) during the convalescent phase, but no virus could be found. The patient recovered completely. Twenty-seven months later, the same patient complained of headaches and was failing his school work. Combined pneumoencephalogram and ventriculogram showed noncommunicating hydrocephalus, consequent to stenosis of the cerebral aqueduct (the aqueduct was dilated rostral to the constriction). A ventriculocardiac shunt was inserted. (5 refs.) - *E. Kravitz.*

Cleveland Metropolitan General Hospital
Cleveland, Ohio 44109

1683 **MEYER, JOHN STERLING; BAUER, RAYMOND B.; RIVERA-OLMOS, VICTOR M.; NOLAN, DAVID C.; & LERNER, A. MARTIN.** *Herpesvirus hominis* encephalitis: Neurological manifestations and use of idoxuridine. *Archives of Neurology*, 23(5):438-450, 1970.

Neurologic details are presented of 8 patients (6 female, 2 male), 10 to 64 years old, with *Herpesvirus hominis* encephalitis; 6 of these patients were treated with idoxuridine, 4 successfully. Symptoms included disturbances of mentation, headache, stupor, coma, obtundity, agitation, confusion, nuchal rigidity, hemiparesis, palsies, convulsions, myoclonus, sensory and motor deficits, cranial nerve paralysis (especially numbers 6 and 7), papilledema, and decorticate and decerebrate posturing. CSF findings varied; EEG findings were abnormal. Intravenous idoxuridine (86 mg/kg/day for 5 days) resulted in remission of

symptoms for at least 18 months. (30 refs.) - *E. Kravitz.*

Baylor College of Medicine
Houston, Texas 77025

1684 SOLOMON, J. B. Unification of foetal and neonatal immunology. *Nature*, 227(5261):895-897, 1970.

The onset of immunologic function is shown to be similar in various species of animals, when age equivalence is employed. The basic point developed was a direct comparison of physiologic age among various birds and mammals. Important parameters include body weight (fetal and postnatal) at peak growth velocity and the first appearance of small lymphocytes. Animals evaluated include chicken, hamster, mouse, rabbit, dog, cattle, monkey, and man. (54 refs.) - *E. Kravitz.*

University of Aberdeen
Aberdeen, Scotland

1685 HORSTMANN, D. M.; LIEBHABER, H.; & KOHORN, E. I. Post-partum vaccination of rubella-susceptible women. *Lancet*, 2(7681):1003-1006, 1970.

Successful post-partum vaccination of women susceptible to rubella is reported. Routine prenatal use of rubella hemagglutination inhibition (HAI) antibody testing is recommended. Vaccination was performed on 74 seronegative women and 31 with low HAI titers (less than 1:8) 2-4 days after delivery; 92% of the seronegative patients developed antibodies, and 16% of those with low titers showed at least 4-fold increases. Joint involvement, the major side-effect after vaccination, occurred in less than 10% of the cases. (12 refs.) - *E. Kravitz.*

Yale University School of Medicine
New Haven, Connecticut

1686 WEAR, DOUGLAS J.; & RAPP, FRED. Encephalitis in newborn hamsters after intracerebral injection of attenuated human measles virus. *Nature*, 227(5265):1347-1348, 1970. (Letter)

Encephalitis developed in newborn hamsters after the intracerebral injection of attenuated measles virus. The titers of infected brain homogenates were higher after injection of attenuated virus than a more virulent strain (Edmonston). Also, the attenuated virus gave signs of clinical measles encephalitis in more animals than did the virulent strain. The need for a more attenuated measles virus, using the hamster brain as a bioassay, is suggested. (11 refs.) - *E. Kravitz.*

Pennsylvania State University
Hershey, Pennsylvania 17033

1687 USATEGUI-GOMEZ, MAGDELANA; HOPKINS, M. S.; & FERNANDEZ DE-CASTRO, AURORA. Serum proteins in amniotic fluids in erythroblastosis fetalis. *Obstetrics and Gynecology*, 36(6):865-874, 1970.

The concentrations of 12 proteins studied in cord blood and sera of 18 Rh negative women remained unaltered throughout pregnancy. However, total proteins in cord blood increased in certain cases; there seemed to be a maternal origin. Individual proteins were determined as a percentage of total protein. (35 refs.) - *E. Kravitz.*

Putnam Memorial Hospital Institute
for Medical Research
Bennington, Vermont

1688 Lead poisoning: Children at risk. *Nature*, 228(5278):1253, 1970.

The problem of lead poisoning among children is of major significance in New York City. Improved testing has resulted in an increase in the number of cases detected, but a decrease in the number of fatalities. Currently, an estimated 6,000-8,000 children in New York City have significant blood lead levels. Extremely high levels can cause irreversible brain damage. Chipping paint in old houses is the primary source of lead. Law enforcement is hampered by insufficient manpower, budget, and knowledge. (No refs.) - *E. Kravitz.*

1689 BERK, PAUL D.; BLOOMER, JOSEPH R.; & BERLIN, NATHANIEL I. Y and Z:

New terms in the bilirubin equation. *New England Journal of Medicine*, 283(21):1166-1167, 1970. (Editorial)

The possible significance of the hepatic intracellular proteins Y and Z in bilirubin metabolism is discussed. Both of these proteins bind organic anions, *in vivo* and *in vitro*, and they are involved with organic anion transfer from plasma to bile. Low intracellular protein seems to account for abnormally high plasma bilirubin and bromsulfalein values during the first 10 days of primate life, but other factors may also be involved. Fetal protein Z seems to reach normal values early. (7 refs.) - *E. Kravitz.*

1690 **GOLDFIELD, MICHAEL; & WEINSTEIN, MORTON R.** Lithium in pregnancy: A review with recommendations. *American Journal of Psychiatry*, 127(7):888-893, 1971.

Lithium is used as a psychopharmaceutical agent during pregnancy in the event that a patient has had serious manic episodes curtailed previously by lithium and then decides to become pregnant or when a woman has become pregnant despite advice that she avoid pregnancy while on a course of lithium. There has been conflicting evidence regarding deleterious effects on the fetus caused by lithium. Out of 45 known infants delivered to women who were taking lithium during pregnancy, 43 were normal, and 2 had congenital abnormalities. This finding does not exceed the usual percentage in all pregnancies, and no clearcut relationship exists between the anomalies and use of the drug. Nevertheless, indications for lithium administration during pregnancy must be examined closely, birth control should be encouraged for women requiring the drug, and careful monitoring of drug dosages and salt balance in the mother must be accomplished. (22 refs.) - *B. J. Grylack.*

Langley Porter Neuropsychiatric Institute
San Francisco, California 94122

1691 **MARTIN, CL.; BABIN, J.-P.; HEHUNSTRE, J.-P.; & SAN JUAN, B.** Les icteres du nouveau-ne par immunisation anti-Rhesus avec bilirubine conjuguee d'emblee (Neonatal jaundice with direct bilirubin treated by anti-Rhesus immunization). *Archives Francaises de Pediatrie*, 27(8):868-869, 1970.

Three cases of early neonatal jaundice with bilirubin either in the conjugated state or of mixed types, resulting from fetomaternal disease of Rhesus system immunization, were studied. An infant born after 8 months gestation at a weight of 2,280 g presented with mucocutaneous greenish jaundice, hyperpigmented urine, ecchymotic purpura of the face and trunk, enormous hepatosplenomegaly, and soft heart sounds testifying to myocardial difficulties. Bilirubin of cord blood was at 85 mg, of which 63 mg was conjugated, blood group was O Rh positive, anemia was intense. The level of bilirubin at 2 hours of birth was 112 mg, of which 97 was conjugated, and at 24 hours was 204 mg, of which 151 was conjugated. The infant died despite transfusions and resuscitation. An infant weighing 3,300 g was born in a less serious state which remained good despite continuous rise of the bilirubin level to 170 mg, of which 160 was conjugated at 24 hours. A transfusion was performed, but jaundice persisted for 3 weeks. Flocculation tests and transaminases were normal, however, and the final course was favorable under medical treatment. The third case was comparable with the second. Neither microbial or viral infectious hepatitis nor congenital metabolic disease was a possible etiology in these cases. Anemia was significantly responsible for an anoxic fetopathy. (No refs.) - *B. J. Grylack.*

No address

1692 **PAPIERNIK, MARTINE; & NEZELOF, CHRISTIAN.** La promotion immunitaire du foetus humain (Immunological promotion of the human fetus). *Archives Francaises de Pediatrie*, 27(8):777-784, 1970.

Immunological mechanisms are under the essential control of the lymphoid system, with the thymus being the earliest and most elaborate of the lymphoid organs in the course of fetal life. Immunoglobulin (Ig) G is transferred across the placenta from mother to fetus. In addition, the fetus has been found to produce its own Ig's. Intrauterine infections stimulate maturation of the fetal immunological system. Regardless of such stimuli, the human fetus also has a definite

basic rate of IgM and IgG production. Knowledge is much less precise in regard to the development of cellular immunity. It appears that maternal cells are rejected by the fetal organism and that the human fetus is capable of protecting itself eventually *in utero* and after birth. Rubeola in its congenital form is an example of a fetomaternal pathogenic relationship. In certain cases, the result is the affliction of the immunological system itself. In an immunological sense, the previously held image of the totally protective gestating mother must be revised. (45 refs.) - *B. J. Grylack.*

Laboratoire de Pathologie pediatrique
I.N.S.E.R.M., U 77
75-Paris 15^{e}, France

1693 **ZAMET, P.; & CHUNGA, F.** Separation by gel filtration and microdetermination of unbound bilirubin. II. Study of sera in icteric newborn infants. *Acta Paediatrica Scandinavica*, 60:33-38, 1971.

Unbound bilirubin was extracted by Sephadex gel filtration in 45 samples of sera drawn from jaundiced neonates and measured by micromethod to determine the possible presence of unbound bilirubin, to measure its levels, and to study its possible correlation with total bilirubinemia. Neither in infants with erythroblastosis fetalis nor in premature infants could any correlation be established between the total bilirubin level and the amount of unbound bilirubin. The incidence of kernicterus in the sample was small, but the data suggest that kernicterus can be observed in spite of low levels of total bilirubin, and that where there is no free bilirubin there will be no risk of kernicterus. Larger series are necessary to establish correlations between free bilirubin level and the occurrence of kernicterus. (31 refs.) - *N. Jarvis.*

Centre de Recherches Biologiques Neonatales
Paris, France

1694 **CHUNGA, F.; & LARDINOIS, R.** Separation by gel filtration and microdetermination of unbound bilirubin. I. *In vitro* albumin and acidosis effects on albumin-bilirubin binding. *Acta Paediatrica Scandinavica*, 60:27-32, 1971.

An extraction technique and diazoreaction method were devised to separate unbound bilirubin from serum and to analyze small serum volumes drawn from neonates. Sephadex gel filtration shows that pure or unbound bilirubin stays at the origin, whereas bound bilirubin migrates with albumin. When albumin and bilirubin are mixed in variable proportions and at different pH's, bilirubin binding becomes higher as albumin proportions increase. For a given albumin-bilirubin mixture, the proportion of protein bound bilirubin increases with the pH. Acidosis increases unbound bilirubin concentration in serum and tissues and unbinds bilirubin already attached to albumin in serum and albumin-bilirubin mixtures in a proportion apparently related to the degree of acidosis. Increasing the albumin concentration compensates for the acidosis effect. These observations emphasize the importance of acidosis in producing kernicterus and the protective effect of albumin. (33 refs.) - *N. Jarvis.*

Centre de Recherches Biologiques Neonatales
Paris, France

1695 **KINTZEL, H.-W.; HINKEL, G. K.; & SCHWARZE, R.** The decrease in the serum bilirubin level in premature infants by orotic acid. *Acta Paediatrica Scandinavica*, 60:1-5, 1971.

Because of the undesirable effects of phenobarbital and phenylbutazone and the ineffectiveness of nikethamide in the prevention of neonatal hyperbilirubinemia, other agents have been sought to prevent a toxic increase of serum bilirubin. Japanese authors investigating the influence of orotic acid on neonatal bilirubin level found it to have a slight effect on bilirubin level in a small number of neonates. In the present investigation, after preliminary experiments with a 100 mg dose, the influence of a daily dose of 300 mg of orotic acid on the serum bilirubin level of 102 premature infants was investigated, an equal number of children serving as control group. The serum level for indirect bilirubin analyzed from the third to the sixth day could be decreased in this manner to a statistically significant extent. Only 4 blood transfusions were required in the group treated with orotic acid, whereas 30 exchange transfusions were required in the control group. Although no deleterious effects were noted in the infants treated,

fatty degenerations of the liver and crystallization of orotic acid in the renal tubuli have been noted in animal studies as effects of larger doses. As the effectiveness of the substance is related to dosage, the authors are initiating studies on the mode of action of orotic acid and the possible dangers of side effects of larger doses in humans. (22 refs.) - *N. Jarvis.*

Paediatric Hospital of the Medical
Academy "Carl Gustav Carus"
Dresden, Democratic Republic of Germany

1696 Can prenatal immunization be inherited? *Medical World News*, 12(12):18-19, 1971.

An experimental antigen was administered to female rats before mating in an attempt to demonstrate a significant immunological reaction in the offspring. This reaction could be meaningful in terms of immunizing mothers with long-acting vaccines in order to protect fetuses from the teratogenic effects of prenatal and postnatal infections. The antigen was composed of glutamic acid, lysine, and tyrosine complexed with methylated bovine serum albumin or polylysine. A challenge dose of the synthetic antigen caused the offspring of immunized mothers to produce 5 times as much antibody as animals not exposed previously to the artificial polymer. The baby rat was shown to be born with circulating antibodies plus the capability of responding vigorously to another dose of the antigen after birth. This finding may be applicable in the future to human conditions in which congenital abnormalities predispose to infection directly after birth or in which congenital anomalies caused by prenatal infection may be prevented entirely. (No refs.) - *B. J. Grylack.*

1697 SEVERI, FRANCESCA; RONDINI, G.; ZAVERIO, SILVIO; & VEGNI, MARIA. Prolonged neonatal hyperbilirubinemia and pregnane-3(α),20(β)-diol in maternal milk. *Helvetica Paediatrica Acta*, 25(5):517-522, 1970.

A prolonged neonatal jaundice related to breast milk was observed in 13 infants over a period of 21 months. In no case were other causes of hyperbilirubinemia demonstrable. A determination of pregnane-3(α),20(α)-diol in maternal milk demonstrated the presence of the steroid in 10 of the 13 cases with the quantity varying from 15 to 45 μg/100 ml. The administration of milk containing pregnane-3(α),20(β)-diol to a non-jaundiced newborn did not cause a rise in serum bilirubin. Pregnane-3(α),20(β)-diol could not be detected in breast milk from 15 mothers of non-jaundiced newborns, serving as controls. (17 refs.) - *P. Custers.*

Pediatric Clinic, University of Pavia
Pavia, Italy

1698 CONNOLLY, J. H.; HAIRE, MARGARET; & HADDEN, DIANA S. M. Measles immunoglobulins in subacute sclerosing panencephalitis. *British Medical Journal*, 1(5739):23-25, 1971.

Measles specific immunoglobulins M and G (IgM and IgG) were found in 3 patients with subacute sclerosing panencephalitis (SSPE). Tests in 10 patients with measles showed a persistence of measles IgG after infection but no detectable IgM 46 days after onset of measles rash. The 3 patients had had measles 11 to 13 years prior to onset of SSPE, but measles IgM was still detectable in high titers in serum and CSF. Measles virus antigen was also demonstrated in the brains of these patients. Immunoglobulins thus appear to be produced within the central nervous system; however, whether this represents a persistent infection or a reinfection is as yet undetermined. (17 refs.) - *E. L. Rowan.*

Queen's University of Belfast
Belfast BT12 6BN, Northern Ireland

1699 PHIBBS, R. H.; HARVIN, D.; JONES, G.; TALBOT, C.; COHEN, M.; CROWTHER, D.; & TOOLEY, W. H. Development of children who had received intra-uterine transfusions. *Pediatrics*, 47(4):689-697, 1971.

Of 66 nonhydropic fetuses who received intrauterine transfusions for severe erythroblastosis fetalis, 24 survived the neonatal period and were followed for from 1 to 5 years in order to assess development. Seventeen had shown cardiorespiratory distress at birth, and all but one had hyperbilirubinemia. One child died with severe

central nervous system damage, but the others showed normal growth and development. Intellectual function appeared generally normal with one of the children in the dull-normal range, 1 mildly MR, and 1 questionable. One child had hearing loss (he also received high doses of kanamycin), and one had a speech defect. It was noted that the development of expressive language skills lagged in many of these children. Intrauterine transfusion combined with aggressive therapy in the neonatal period is compatible with grossly normal neurological development. (9 refs.) - *E. L. Rowan.*

University of California, San Francisco
San Francisco, California 94122

1700 **HORTA-BARBOSA, LUIZ; KREBS, HELENE; LEY, ANITA; CHEN, TA-CHUAN; GILKESON, MARY R.; & SEVER, JOHN L.** Progressive increase in cerebrospinal fluid measles antibody levels in subacute sclerosing panencephalitis. *Pediatrics*, 47(4):782-783, 1971.

Measles complement-fixing and hemagglutination inhibition antibodies in both serum and spinal fluid were found and were noted to increase during the course of subacute sclerosing panencephalitis (SSPE) in 25 patients. There was no measles antibody detected in spinal fluid of controls (5 patients with multiple sclerosis and 2 with Jakob-Creutzfeldt disease), and in none was the serum antibody titer higher than 64. The antibody determination can be of importance in the diagnosis of SSPE. (3 refs.) - *E. L. Rowan.*

National Institutes of Health
Bethesda, Maryland 20014

1701 **MILLER, LOUIS H.; REIFSNYDER, DAVID N.; & MARTINEZ, S. ALICIA.** Late onset of disease in congenital toxoplasmosis. *Clinical Pediatrics*, 10(2):78-80, 1971.

Infants infected *in utero* with *Toxoplasma gondii* may be born with encephalomyelitis, hydrocephalus, retinochoroiditis, or hemolytic anemia in all combinations. Contrary to widespread belief, there are usually no signs of disease in the neonatal period. Later, however, these infants may develop hydrocephalus, retinochoroiditis, seizures, or MR. In the case of identical twin boys reported here, congenital toxoplasmosis was suspected following the recognition of hydrocephalus in one twin 9 months after a normal but premature birth. Bilateral peripheral retinochoroiditis was found funduscopically; the physical examination was otherwise normal except for symmetrical hyperreflexia of the patellar reflexes and bilateral Babinski signs. The diagnosis of congenital toxoplasmosis was confirmed by the high Toxoplasma titers in the mother, in the baby, and in the other twin in whom hydrocephalus had developed at 3½ months. Although retinochoroiditis may have been present from birth, both infants had developed normally and were asymptomatic until the hydrocephalus appeared. Both babies were treated for 3 weeks with pyrimethamine, triple sulfonamides, and folinic acid. Since discharge from the hospital the twin who became hydrocephalic at 9 months has shown no evidence of psychomotor retardation and was able to walk by 16 months. His brother lagged behind in growth and development and is beginning to walk at 20 months. With the Sabin-Feldman dye test or the indirect fluorescent antibody test a high titer at birth which persists for 3 or 4 months along with a high titer in the mother is diagnostic of congenital toxoplasmosis. Retinochoroiditis may be the only evidence of a latent infection or may be accompanied by other sequelae of toxoplasmosis such as intracerebral calcification and seizures. Later in childhood the diagnosis of congenital toxoplasmosis becomes more difficult, since a positive serology finding may be the result of an acquired infection. (13 refs.) - *J. C. Moody.*

Columbia University
College of Physicians and Surgeons
New York, New York 10032

1702 Jaundice of the newborn and phenobarbitone. *Lancet*, 1(7690):119-120, 1971. (Editorial)

Phenobarbitone given to expectant mothers in the last week or two of pregnancy can significantly reduce the peak plasma-bilirubin levels in the newborn and lower the incidence of severe hyperbilirubinemia. The basis for this action of phenobarbitone is believed to be linked to its ability to increase glucuronyl-transferase activity in the liver of the fetus or neonate. Treatment of

only the newborn is less effective, and combined treatment of both mother and child seems to give the best results. The wisdom of using phenobarbitone either for routine prophylaxis or in special situations to guard against the potential effects of hyperbilirubinemia has been hotly debated. While there is little doubt that very high levels of plasma-unconjugated bilirubin (above 20 mg/100 ml) may cause severe kernicterus, it is less clear what risk there is from lower plasma-bilirubin levels. There are conflicting answers to the question whether the use of an "enzyme-inducer" such as phenobarbitone in late fetal and early neonatal life produces changes in enzyme activity and differentiation that could have serious long-term effects. Although the evidence concerning possible harmful effects of phenobarbitone in the perinatal period is fragmentary and largely theoretical, there is a good case for its use as a routine prophylactic measure in populations with a high incidence of severe hyperbilirubinemia where the threat of kernicterus outweighs any potential danger. Limited studies of phenobarbitone therapy in infants with rhesus incompatibility indicate that it is also beneficial in the prophylaxis of rhesus hemolytic disease. More data are required, however, concerning the extent of bilirubin toxicity in the newborn and the long-term effects of phenobarbitone therapy in the perinatal period. In addition, the role of other bilirubin-lowering techniques such as phototherapy must be more fully assessed before the prevention of damage by hyperbilirubinemia in the newborn can be attempted on a rational basis. (24 refs.) - *J. C. Moody.*

1703 SILVERMAN, JOSEPH A.; WINTERS, ROBERT W.; & STRANDE, CARL. Lithium carbonate therapy during pregnancy: Apparent lack of effect upon the fetus. *American Journal of Obstetrics and Gynecology*, 109(6):934-936, 1971.

A 23-year-old Caucasian nulliparous Latvian woman received daily dosages of 900 to 1200 mg lithium carbonate for treatment of her uncontrollable manic behavior. While on the lithium carbonate treatment she became pregnant, but this was not recognized until the seventh month. The drug was continued at the same dosage until term. Labor lasted less than 6 hours. The female infant born after approximately 6 hours' labor weighed 3,290g and had Apgar scores of 6, 6, and 9 at 1, 2, and 5 minutes. Physical examination was entirely unremarkable at birth, except for minimal generalized hypotonia which lasted 48 hours. Daily examinations revealed no other clinical abnormalities. Serial electrocardiograms, electroencephalograms, and neuromuscular conduction studies likewise showed no abnormalities. Serial measurements of plasma lithium concentration determined by atomic absorption spectrophotometry in the infant carried out over the first 10 days showed that the lithium completely cleared from her blood in an exponential manner. It would appear from this patient and from Scandinavian literature noting overt congenital malformations in only 2 out of 40 infants of lithium-treated mothers, that lithium carbonate has no apparent teratologic effects; nor does it seem to have any measurable influence upon the ability of the newborn to make a satisfactory adjustment to extrauterine life. The relationship of lithium therapy to birth defects in the 2 Scandinavian infants reported remains to be demonstrated. (3 refs.) - *J. C. Moody.*

Babies Hospital
New York, New York 10032

1704 MIRKIN, BERNARD L. Placental transfer and neonatal elimination of diphenylhydantoin. *American Journal of Obstetrics and Gynecology*, 109(6):930-933, 1971.

The transplacental passage of diphenylhydantoin has been demonstrated in 3 pregnant epileptic females receiving standard oral therapeutic doses of the anticonvulsant (300 mg per day). Maternal plasma concentrations ranged from 3.5 to 6.3 μg per ml, whereas fetal and maternal levels in a given subject were virtually identical at term. This suggests attainment of an equilibrium state in mothers receiving long-term anticonvulsant therapy. Clearance of diphenylhydantoin from neonatal plasma required about 72 hours with a half-life between 50 and 60 hours. The slow elimination of diphenylhydantoin in the newborn probably reflects both hepatic and renal immaturity. Cleft lip was present as an isolated anomaly in 2 of the 3 cases reported. The remaining infant had a cleft palate and lip. A prospective study is required to validly assess the teratologic potentiality of administering this agent during human gestation. (9 refs.) - *J. C. Moody.*

University of Minnesota College of Medicine
Minneapolis, Minnesota

1705 **MARCUSE, EDGAR K.; PETERSON, DONALD R.; THOMPSON, ROBERT S.; & CHINN, NINA.** Retrospective survey of congenital rubella syndrome in the State of Washington, 1963-68. *HSMHA Health Reports*, 86(2):161-167, 1971.

A retrospective survey of congenital rubella syndrome (CRS) demonstrated the feasibility of such surveillance and suggested possible surveillance methods. The study, conducted between 1963-68, set certain CRS criteria which were met by 126 Ss. Sources of case identification information were birth and death certificates; hospital reports; referral center records of speech, hearing, and congenital defects; and reports from physicians, special education teachers, and parents. The fact that the reports from special education teachers and the referral center accounted for 119 cases suggests that surveillance be concentrated on these sources in urban areas. Reporting in rural areas should be the responsibility of physicians. Analysis of the cases indicated that the defects identified did not represent the true incidence of defects due to fetal infection. More rigorous and accurate reporting criteria will be possible with more widely available laboratory facilities for rubella serologic testing and greater understanding of the disease as well as the importance of reporting it. (4 refs.) - *S. Glinsky.*

Washington State Division of Health
Olympia, Washington 98501

1706 Medical aspects of childhood lead poisoning. *HSMHA Health Reports*, 86(2):140-143, 1971.

Guidelines are enunciated by the Public Health Service for the development of screening programs to identify, treat, and control lead poisoning and absorption in children 1 to 6 years old. All children living in or exposed to environments or housing utilizing lead-based paints must be screened. Plumbism is present when blood levels of lead exceed 40μg per 100 ml of whole blood. Children with over 80μg have unequivocal cases of lead poisoning and must be hospitalized and given therapy due to the risk of encephalopathy, which in 40% of cases leads to severe and permanent brain damage. Those with blood lead values of 50 to 79μg who are not diagnosed as having lead poisoning should have blood lead values determined at monthly intervals; this is particularly important during the summer months. Children with blood lead values of 40 to 49μg who are exposed to lead hazards in their homes should be examined for level of lead in the blood every 6 to 8 weeks. In addition to treatment and follow-up to the age of 6 years in all cases of plumbism, neurological, psychological, clinical and laboratory evaluations are to be recorded. Above all the sources of lead poisoning should be removed; this is the responsibility of local government. (16 refs.) - *S. Glinsky.*

1707 **MURPHREE, ODDIST D.; MORGAN, PAUL N.; & JARMAN, RUTH V.** Learning deficits and activity changes: A partial laboratory model in postencephalitic rats for studies of brain damage. *Conditional Reflex*, 6(1):30-35, 1971.

The rat could be an appropriate model for studies of postencephalitic behavioral changes caused by viral agents when central nervous system damage is the area of interest and when learning and activity behaviors are measured. Sprague-Dawley rats ranging from 18 to 21 days of age were inoculated in the right cerebral hemisphere with St. Louis, Japanese B, and Murray Valley encephalitis virus. Mumps virus was injected at age 1 day. Very young injected rats developed fatal encephalitis while rats injected at ages of more than 2 weeks showed little or no clinical signs of infection. Inoculations between these extremes resulted in clinical disease in most rats, with many surviving. Physically most of these surviving animals appeared normal, but when tested at maturity in a rather complex maze, they showed a marked reduction in learning ability. Tilting-cage-type activity measurements taken overnight with the Murray Valley encephalitis group also indicated that most of the postinfectious rats were hyperactive. Such a laboratory model might be related to similar hyperactivity and learning disabilities seen in certain types of minimally brain damaged children. (8 refs.) - *S. Glinsky.*

Veteran's Administration Hospital
Little Rock, Arkansas

1708 **MEYER, HARRY M., JR.; & PARKMAN, PAUL D.** Rubella vaccination, a review of practical experience. *Journal of the*

American Medical Association, 215(4):613-619, 1971.

The results of rubella virus vaccine use between June 10, 1969, and October 1, 1970, in the United States confirm on the whole both the safety and efficacy of the vaccine. During this period approximately 28 million doses were distributed, and the experience since licensure has reinforced earlier data regarding vaccine safety. Troublesome reactions clearly associated with immunization have largely been confined to rubella-like symptoms of joint pain and related complaints. Community-wide vaccine use has not posed a significant risk of attenuated virus transmission to contacts. The inadvertent inoculation of pregnant women represents the greatest preventable immunization problem. New studies give additional information regarding the quality and durability of vaccine-induced immunity. Although there exists some disagreement concerning the interpretation of these new data, the present recommendations for vaccine use represent the best of options available for preventing maternal-fetal rubella in the United States. (36 refs.) - *S. Glinsky.*

National Institutes of Health
Bethesda, Maryland

1709 SCOTT, H. DENMAN; & BYRNE, EARL B. Exposure of susceptible pregnant women to rubella vaccinees. *Journal of the American Medical Association*, 215(4):609-612, 1971.

Susceptible pregnant women exposed to rubella vaccine virus are as unlikely to acquire infection as are susceptible siblings or nonpregnant women. In a state-wide rubella vaccination program, where 75,000 children were vaccinated, 1,350 pregnant women were tested serologically before and during the 2 month campaign. Of these only 13% were identified as susceptible. Follow-up serological studies on 121 of this susceptible group, which included 31 close contacts with vaccinees, revealed only one case of seroconversion. This clinically asymptomatic patient could not be related temporally to an immunized child in the home or to any other identifiable exposure. The results reinforce the view that there is no reason to suspect that pregnant women are more predisposed to vaccine virus infection than are siblings or nonpregnant mothers of vaccinees. (6 refs.) - *S. Glinsky.*

Rhode Island Department of Health
Providence, Rhode Island

1710 CHIN, JAMES; EBBIN, ALLAN J.; WILSON, MIRIAM G.; & LENNETTE, EDWIN H. Avoidance of rubella immunization of women during or shortly before pregnancy. *Journal of the American Medical Association*, 215(4):632-634, 1971.

Several cases of inadvertent inoculation indicate that further precautions must be taken to prevent the administration of rubella vaccines to women either during pregnancy or at least 2 months before pregnancy. Seventeen such cases occurred in California between April 1969 and February 1970. Eleven of the women, tested for hemagglutination inhibiting antibody after immunization, possessed demonstrable rubella antibody. However, it is not possible to decide whether the antibody was a vaccine response or a reflection of a prior natural infection with the virus. Seven of the pregnancies were terminated, 4 resulted in normal infants, and the remainder have not gone full term. These cases point up the necessity for close and frank consultation between physician and patient regarding possible rubella side effects and careful record-keeping regarding vaccine administration. (4 refs.) - *S. Glinsky.*

California State Department of Public Health
Berkeley, California

1711 MONIF, GILLES R. G.; & HELD, BEREL. Noncontagiousness of the Cendehill vaccine strain of rubella virus from mother to newborn infant. *Journal of Pediatrics*, 78(2):306-307, 1971.

There was no evidence of rubella infection in the progeny of 10 women vaccinated with a rubella virus in the postpartum period. Prior to the fourth postpartum day an obstetric population was administered the Cendehill vaccine strain of rubella virus and 10 mothers developed seroconversions. No antibody to rubella virus was identified in the sera of any of the infants of these mothers. The infants were followed from 6 to 20 weeks. The results indicate that the HPV-77 and Cendehill vaccine strains of rubella virus are not

readily contagious under normal circumstances. (3 refs.) - *S. Glinsky.*

University of Florida College of Medicine
Gainesville, Florida

1712 **NELSON, MATILDA M.; & FORFAR, JOHN O.** Associations between drugs administered during pregnancy and congenital abnormalities of the fetus. *British Medical Journal*, 1(5748):523-527, 1971.

As part of a survey of the etiology of congenital malformations, the consumption of prescribed and self-administered drugs in 1,369 pregnancies was examined retrospectively. Ninety-seven percent of the women took prescribed drugs at some time during their pregnancy, and 65% took self-administered drugs. Of the 458 mothers who gave birth to infants with congenital abnormalities, a significantly greater number took aspirin, antacids, dextroamphetamine, phenobarbital, sodium amytal, other barbiturates, cough medications, iron, sulfonamides, and nicotinamide as compared with mothers who did not give birth to infants with congenital disorders. However, since the majority of mothers taking analgesics, antacids, appetite suppressants, barbiturates, cough medicines, iron, sulfonamides, and vitamins produced normal babies, the teratogenic potency of these drugs must be low. Drugs should be prescribed to pregnant women and to women in whom conception is likely with extreme caution. Under the same circumstances, self-medication should be avoided. (21 refs.) - *B. J. Grylack.*

University of Edinburgh
Edinburgh, Scotland

1713 **WILLOUGHBY, H. W.; DESJARDINS, P. D.; POWER, R.M.H.; & LEE, T. K.** Hyperbilirubinemia in late pregnancy and its correlation with meconium staining (symposium). *American Journal of Obstetrics and Gynecology*, 109(3):383-388, 1971.

To establish the importance of hyperbilirubinemia in the last trimester of pregnancy as a prognostic sign, the incidence of this condition in the patient population and the correlation of the abnormality with meconium staining and fetal distress during delivery and with neonatal morbidity rates and perinatal mortality rates were studied. The incidence of hyperbilirubinemia among 1,018 pregnant women was 10.5% and the incidence of meconium staining was 15.8% in the same group. The frequency of meconium staining during delivery either in the amniotic fluid or on the baby or both is much higher in hyperbilirubinemia patients than in women with normal serum bilirubin levels. The incidence of fetal distress was 24.0% in all patients with meconium staining and 2.9% in patients without it. Definite correlations could not be drawn from figures obtained on neonatal and perinatal morbidity and mortality. Testing for hyperbilirubinemia should be included in routine laboratory blood tests carried out on pregnant women during the last trimester of pregnancy, and patients with hyperbilirubinemia should be considered pregnancies at risk. (14 refs.) - *B. J. Grylack.*

McGill University
Montreal, Quebec, Canada

1714 **PRITCHARD, JACK A.; SCOTT, DANIEL E.; & WHALLEY, PEGGY J.** Maternal folate deficiency and pregnancy wastage. IV. Effects of folic acid supplements, anticonvulsants, and oral contraceptives (symposium). *American Journal of Obstetrics and Gynecology*, 109(3):341-346, 1971.

The hematologic benefits from routine supplementation with iron plus folic acid as compared to iron alone, the effects on the products of conception of maternal folate deficiency induced by anticonvulsant drugs, and the effects of oral contraceptives on folate metabolism in the nonpregnant women were evaluated in a study of the relationship between maternal folate deficiency and reproductive failure. The results showed that folic acid supplementation did not have value in a group of 25 women, whereas administration of iron had a definite effect on the maternal hemoglobin level. Observations of 77 pregnancies in 43 epileptic women receiving anticonvulsants demonstrated that the increased frequency of maternal folate deficiency which was caused by ingestion of diphenylhydantoin and related drugs throughout pregnancy was not deleterious to the fetus. Observations further revealed that plasma folate

levels were not lower in women who had taken oral contraceptives for a prolonged period. In otherwise untreated patients with megaloblastic anemia, an oral contraceptive in large dosage did not block the response to the folate in an ordinary hospital diet. Continued use of oral contraceptives by these women has not resulted in any evidence of hematologic relapse. (29 refs.) - *B. J. Grylack.*

University of Texas (Southwestern) Medical School
Dallas, Texas

1715 SAYRE, JAMES W.; & WILSON, DAVID J. A spot test for detection of lead in paint. *Pediatrics*, 46(5):783-785, 1970.

A simple spot test requiring little technical ability for its use affords a quick and reliable method for detecting lead in paint. A 5 to 8% solution of sodium sulfide in a 5 ml polyethylene squeeze dropper bottle, when applied to paint chips or scrapings, can qualitatively detect lead in paint at levels as low as 0.8%. The formation of a black precipitate of lead sulfide is the basis of this test. Other metals which also form black sulfides are usually present in paint at levels of less than 1%, whereas paint may contain as high as 50 to 70% of lead. The procedure requires little training for its use and interpretation and can easily be utilized in the home, particularly where children are known to pick at the walls or windowsills. The test should prove useful in screening campaigns to identify early cases of lead poisoning. (5 refs.) - *M. S. Fish.*

University of Rochester School of Dentistry
Rochester, New York 14260

1716 RAHAL, JAMES J., JR.; & HENLE, GERTRUDE. Infectious mononucleosis and Reye's syndrome: A fatal case with studies for Epstein-Barr virus. *Pediatrics*, 46(5):776-780, 1970.

A high titer of antibody to Epstein-Barr virus (EBV) was found in the serum of a patient with infectious mononucleosis and Reye's syndrome, although impression smears of affected organs failed to demonstrate antigens of EBV. The S, a 6-year-old male, developed toxic encephalitis and severe hepatitis following the onset of infectious mononucleosis. Despite treatment with oral glycerol and parenteral urea, mannitol, dexamethasone, penicillin, and streptomycin, and the use of a mechanical ventilator, the patient did not respond and died following cardiac arrest. Serum specimens taken on consecutive days gave titers of 1:160 by the indirect immunofluorescent test for antibodies to EBV. Although autopsy showed brain edema, fatty necrosis of the liver, and a mononuclear infiltration of the liver, spleen, lymph nodes, and gastrointestinal tract, neither antigens of EBV nor cytopathic effect could be demonstrated in smears or suspensions of these organs. (25 refs.) - *M. S. Fish.*

New York (Manhattan) Veterans
Administration Hospital
New York, New York 10010

1717 MAREK, JOSEF; & HORKY, KAREL. Aminoglutethimide administration in pregnancy. *Lancet*, 2(7686):1312-1313, 1970. (Letter)

Idiopathic posture-dependent edema was diagnosed in a 27-year-old female patient, and the administered combination of aminoglutethimide and furosemide produced desirable diuretic and natriuretic effects. Aminoglutethimide was stopped when gynecological examination revealed a 3-month pregnancy because of previous reports of unfavorable effects of the drug on gonadal and reproductive function in women. The patient gave birth to a healthy girl with no signs of virilization, and after 2 years there were no signs of abnormal somatic or psychological development. This case shows that aminoglutethimide does not inhibit ovulation or produce sterility in women as it does in rats. Although the administration of aminoglutethimide here did not cause miscarriage or defects in the child, the drug should be used with extreme caution in fertile women and should be discontinued as soon as pregnancy is suspected. (8 refs.) - *B. J. Grylack.*

Charles University
Prague, Czechoslovakia

1718 Prevention de la rubeole congenitale (Prevention of congenital rubella). *Amentia*, 23:14-15, 1971.

The incidence of congenital rubella in nonepidemic periods is (according to Sever) 1 to 2 cases

per 1,000 pregnancies, corresponding to approximately 300 cases annually in Belgium. The risk of malformations for the child varies according to the time of gestation, with from 60 - 90% of malformations occurring in the first month, and only 5% from the third month on. Cooper recorded, that among 376 children born of mothers who had contracted rubella in early pregnancy, 45.2% show psychomotor retardation, while 67% have different degrees of deafness. In several retrospective studies, congenital rubella has been implicated in 0.5 to 5% of all cases of MR. To prevent the occurrence of congenital rubella and the subsequent dangers of MR for children, the systematic vaccination of all teenage girls is recommended as an immediate measure. (No refs.) - *P. Custers.*

1719 **KAPITULNIK, J.; SCHENKER, J. G.; & BLONDHEIM, S. H.** Diagnosis of fetal hemolytic disease by a chemical test in a case with metheme-contaminated amniotic fluid. *American Journal of Obstetrics and Gynecology*, 110(1):62-64, 1971.

Contamination of amniotic fluid with metheme pigments interferes with the spectrophotometric estimation of bilirubin, decreasing the ΔOD_{450}, but it has no effect on the chemical determination of bilirubin by the diazo reaction. The brown-tinted amniotic fluid of a 30-year-old, type A, Rh-negative mother, after artificial insemination with Rh-negative donor semen, was shown to contain metheme pigments. Although the spectrophotometric evaluation (ΔOD_{450}) of the contaminated fluid was normal, the chemically determined bilirubin concentration correctly indicated the presence of hemolytic disease of the fetus, which was later shown to be Rh-positive. The results show that sole reliance on the spectrophotometric method may lead to underestimation of the severity of hemolytic disease of the fetus or a missed diagnosis. (6 refs.) - *S. Glinsky.*

Hadassah-University Hospital
Jerusalem, Israel

1720 **QUEENAN, JOHN T.; GADOW, ENRIQUE C.; & LOPES, ANTONIO C.** Role of spontaneous abortion in Rh immunization. *American Journal of Obstetrics and Gynecology*, 110(1):128-130, 1971.

The role of spontaneous abortion in Rh immunization is minimal. Of 98 Rh-negative and previously unimmunized patients who had 165 spontaneous abortions, only 3 women were found to have anti-Rh antibodies. A corrected incidence of maternal Rh immunization for this group was 3.1%. The incidence of maternal Rh immunization following therapeutic abortion would be even greater due to fetoplacental circulation functioning at the time of pregnancy interruption. Rh immune prophylaxis may be of benefit to some Rh-negative women having spontaneous abortions. (8 refs.) - *S. Glinsky.*

1 Perryridge Road
Greenwich, Connecticut 06830

1721 **KAISER, I. H.** Ballantyne and triple edema. *American Journal of Obstetrics and Gynecology*, 110(1):115-120, 1971.

The nomenclature of triple edema is suggested to describe the syndrome of edema of the mother, placenta, and fetus. Little information regarding hydrops fetalis has been forthcoming since Ballantyne's treatises, except for the identification of the role of Rh isoimmunization in the pathogenesis of some cases, and a more detailed appreciation of placental pathology. A case report of a 24-year-old primipara is presented. The S exhibited edema in the legs and lower back and was delivered by cesarean section of a massively edematous 3,640g male infant, who never achieved respiration. The 1,020g placenta was also edematous. The patient recovered and has since given birth to a normal male infant. At present there is little information on the syndrome of hydrops fetalis, except for an awareness of processes which produce massive fetal edema and which are either incompatible with fetal life or, as in the present case, are hazardous to the fetus only at and after birth. A systematic study of the placentas associated with the triple edema syndrome for evidence of a common metabolic or morphologic lesion is urgently needed. (41 refs.) - *S. Glinsky.*

525 East 68th Street
New York, New York 10021

1722 **BORNSTEIN, MYER S.** Severity of hemolytic disease of the newborn. *Obstetrics and Gynecology*, 37(1):159, 1971. (Letter)

Comparison of the Ovenstone Factor with the $OD_{450}\mu$ method suggests that the former is a highly accurate predictor of the severity of hemolytic disease of the newborn. The results of a study comparing the Ovenstone Factor with the $OD_{450}\mu$ methods yielded an accuracy of 90%. Where a spectrophotometer is not available to read the needed wavelengths, a Coleman Junior spectrophotometer can be used and the OD of 480 and 500μ are obtained by subsequently applying the Ovenstone Formula. This is a useful procedure when facilities are limited. (1 ref.) - *S. Glinsky.*

4609th United States Air Force Hospital
Kincheloe Air Force Base, Michigan 49788

1723 **O'DONOGHUE, S. E. FIDELMA.** Distribution of pethidine and chlorpromazine in maternal, foetal and neonatal biological fluids. *Nature*, 229(5280):124-125, 1971.

The feto-maternal system is permeable not only to pethidine and chlorpromazine, but to many of their metabolites. 220 women received 50 to 100 mg intramuscularly of pethidine, chlorpromazine, or promazine, singly or in combination, shortly before delivery. Maternal, fetal, and neonatal fluids were investigated for these drugs and their metabolites. Pethidine, norpethidine, chlorpromazine and many metabolites were found in the maternal, fetal, and neonatal fluids in varying distributions. To elucidate the origin of metabolites in the urine of the neonate, neonates from untreated mothers received 1 mg doses of pethidine, chlorpromazine, or promazine. Examination of the neonates' urine clearly demonstrated that the newborn can metabolize drugs within 24 hours after birth. A comparison of the 2 groups of neonates showed metabolites in the neonates from treated mothers not found in the neonates from untreated mothers. These results have therapeutic and toxicological implications for the clinician. (7 refs.) - *S. Glinsky.*

The Rotunda Hospital
Dublin, Ireland

1724 **BOYER, SAMUEL H.; NOYES, ANDREA N.; VRABLIK, GEORGE R.; DONALDSON, LOIS J.; SCHAEFER, EDWARD W., JR.; GRAY, CLINTON W.; & THURMON, THEODORE F.** Silent hemoglobin alpha genes in apes: Potential source of thalassemia. *Science*, 171(3967):182-185, 1971.

Small quantities of unusual hemoglobins, which contain unique α chains that differ from ordinary by 8 to 9 scattered amino acid changes, were found in 1 of 37 chimpanzees and in 2 of 6 gorillas. A previously undetected hemoglobin $^3\alpha$ locus is responsible for the unusual chains. Since most apes do not have $^3\alpha$ products, $^3\alpha$ is thought to be synthetically inactive in all but a few reversion mutants. Data supporting the theory that the inactive $^3\alpha$ locus is juxtaposed to an active locus, coupled with the supposition that $^3\alpha$ exists in man, suggest that thalassemia may be caused by nonhomologous recombination between 2 loci. (23 refs.) - *J. K. Wyatt.*

Johns Hopkins University School of Medicine
Baltimore, Maryland 21205

1725 **ZAUSMER, ELIZABETH.** Congenital rubella: Pathogenesis of motor deficits. *Pediatrics*, 47(1):16-26, 1971.

The character and degree of motor deficits in 43 children (CA 3-4½ yrs) with major hearing loss and history of maternal rubella were related to the maternal gestational age at which the infection occurred. Perceptual-motor behavior was evaluated in the areas of muscle tone and strength, flexibility, balance, gross and fine motor skills, ball skills, imitation of gestures, self-care, character of movement, and adaptive behavior. The most severe multiple handicaps were found in children of mothers who had rubella infection during the first 4 weeks of pregnancy. Risk of motor deficit seemed considerably reduced when maternal infection occurred after the eighth week of pregnancy. Children whose mothers had rubella infection between the fifth and eighth week of gestation had the highest incidence of equilibrium defects. (36 refs.) - *J. K. Wyatt.*

Children's Hospital Medical Center
Boston, Massachusetts 02115

1726 **GIUNTA, FRANK.** A 1-year experience with phototherapy for jaundice of prematurity. *Pediatrics*, 47(1):123-125, 1971.

Serum bilirubin was maintained at less than 12 mg/100 ml in 96.9% of 416 premature infants exposed from birth to environmental lighting of 90 footcandles and placed under "daylight" illumination of 500 footcandles when the direct bilirubin approached or rose above 10 mg/100 ml. In a control group of 47 premature infants who were treated from birth and for 6 days under environmental lighting (90 footcandles), serum bilirubin levels remained at 12 mg/100 ml in only 72.5%. Between group differences were significant ($p<.001$). The use of nursery lighting and management of premature infants with rising bilirubin avoided unnecessary exposure of over 80% of these infants to high-intensity illumination. (7 refs.) - *J. K. Wyatt.*

Providence Lying-In Hospital
Providence, Rhode Island 02908

1727 **ROTHSCHILD, EDMUND O.** Lead poisoning—the silent epidemic. *New England Journal of Medicine*, 283(13):704-705, 1970. (Editorial)

Problems associated with lead poisoning are discussed. Blood levels are important diagnostic aids, e.g., 40-60 micrograms/100 grams is suspicious, greater than 60 micrograms is diagnostic. A good microanalytical method is needed. The use of chelation therapy in acute cases requires good judgment; criteria are minimal. Sources of lead poisoning include earthenware and old lead paint. The latter source is particularly common for the large number of child victims (5-20% in certain urban areas), affected via ingestion; mental retardation and institutionalization are among the costly human and financial sequelae. Unfortunately, adequate preventive efforts are still lacking. (4 refs.) - *E. Kravitz.*

No address

1728 Rubella program questioned, but defenders rally. *Medical World News*, 11(34):4, 1970.

Several points of view concerning the rubella immunization program are summarized. Dr. Enders pointed out that a vaccinated female child may lose her immunity by the time she reaches her child-bearing years. Nevertheless, others have stated that a high degree of rubella control seems attainable by vaccinating children en masse. Some degree of reinfection may occur even after naturally acquired rubella. (No refs.) - *E. Kravitz.*

No address

1729 **RUBEN, R. J.** Screening for rubella during pregnancy. *New England Journal of Medicine*, 283(23):1292, 1970. (Letter)

The suggestion is made that all pregnant women should be tested for rubella antibody titers as soon as they become pregnant and at 3 and 6 months. This should detect the presence of subclinical rubella and permit appropriate preventive measures to be taken against fetal damage. The lack of a highly effective vaccine against rubella points up the need for such a routine testing program. (1 ref.) - *E. Kravitz.*

Albert Einstein College of Medicine
Bronx, New York

1730 **HORSTMANN, DOROTHY M.** Screening for rubella during pregnancy. *New England Journal of Medicine*, 283(23):1292, 1970. (Letter)

The suggestion that rubella titers be obtained on pregnant women is worthwhile in order to identify those who are susceptible; vaccination can then be performed post partum (before hospital discharge). However, there is no value to testing at 3 or 6 months of gestation since the 20-30% risk of intrauterine fetal damage occurs during the first trimester. (2 refs.) - *E. Kravitz.*

Yale University School of Medicine
New Haven, Connecticut

1731 **WHITFIELD, C. R.; LAPPIN, T.R.J.; & CARSON, MAX.** Further development and experience in an action line method for the management of rhesus isoimmunization. *Journal of Obstetrics and Gynaecology of the British Commonwealth*, 77(9):791-795, 1970.

During a 30-month period, the action line method of determining the time for intrauterine transfusion or delivery in rhesus iso-immunization was used in 407 pregnancies, and other methods were employed in 127 cases. The action line method is based upon extrapolation of the trend between separate estimations of the amniotic fluid bilirubin pigments, measured by their optical density "peak" at 450 to an "action line" in the third trimester. The rhesus mortality rate in all pregnancies managed correctly by the action line or the other criteria was 11.6%, with a total wastage, including abortions, from all causes of 15.2%, as compared with rhesus mortality of 31.0% in pregnancies that were managed incorrectly during the same period. The action line is an approximate and easily interpreted method of timing intervention in about 4/5 of rhesus incompatibility cases. (7 refs.) - *B. J. Grylack.*

Royal Maternity Hospital
Belfast, Northern Ireland

1732 LEWAK, NORMAN. Measles (rubeola) and rubella: (1) Dangers of the layman's confusion. *Pediatrics*, 46(6):978, 1970.

Due to the apathy of the public and the medical profession in regard to the danger represented by measles, unnecessary morbidity and mortality have occurred which could have been prevented by a mass immunization campaign. Inasmuch as lay persons are confused about the distinction between rubella and rubeola, a large-scale publicity campaign should be initiated in order to differentiate rubeola from rubella from other exanthemata and to offer the reminder that rubeola can cause morbidity and mortality. (No refs.) - *B. J. Grylack.*

2219 Santa Clara
Alameda, California 94501

1733 KATZ, SAMUEL L. Measles (rubeola) and rubella: (2) Mass measles immunization. *Pediatrics*, 46(6):978-979, 1970.

Considerable confusion exists in regard to the distinction between rubeola and rubella. Since 1966 the Committee on Infectious Diseases of the American Academy of Pediatrics has attempted to maintain a firm policy in all written material of designating the 2 diseases as measles and rubella. It is not true that no mass measles immunization has ever been conducted. In the first 6 years following the licensure of measles virus vaccine in 1963, more than 35,000,000 doses were distributed in the United States in various health care delivery settings. Measles vaccine must continue to be included among the immunizations scheduled for all infants in the first 2 years of life. (No refs.) - *B. J. Grylack.*

Duke University Medical Center
Durham, North Carolina 27706

1734 GREENBERG, ROBERT E.; & CHRISTIANSEN, ROBERT O. The critically ill child: Hypoglycemia. *Pediatrics*, 46(6):915-920, 1970.

Hypoglycemia represents a significant emergency situation in pediatrics, primarily because of the deleterious neurophysiologic sequelae of glucose deprivation on the central nervous system. Widely accepted diagnostic criteria of hypoglycemia include 2 or more blood glucose values of less than 30 mg/100 ml in full-term infants, concentration of blood glucose of less than 20 mg/100 ml in the neonate weighing less than 2,500 g, and blood glucose values of less than 40 mg/100 ml in the older infant and child. Two major predisposing factors to neonatal hypoglycemia are placental dysfunction and postnatal illness. Glucose, hydrocortisone, and glucagon are parenteral agents of treatment in neonatal hypoglycemia. Permanent neurologic sequelae result with greater frequency when repetitive episodes of hypoglycemia occur prior to 6 months of age. Recognition of hypoglycemia in the older infant and child is easier than in the newborn, and glucose is the only agent needed for treatment of an acute episode in older children. Concentration of blood glucose is essentially a result of 2 interacting processes, peripheral glucose utilization and hepatic glucose production. Whenever possible, therapy for hypoglycemia should be directed towards causative mechanisms. (13 refs.) - *B. J. Grylack.*

Stanford University School of Medicine
Stanford, California 94305

1735 GUTHRIE, RICHARD; VAN LEEUWEN, G.; & GLENN, LINDA. The frequency of

asymptomatic hypoglycemia in high risk newborn infants. *Pediatrics*, 46(6):933-936, 1970.

One hundred and nine high risk infants and 25 normal term infants were studied to assess the frequency of low blood glucose levels in a high risk asymptomatic group of infants. The high risk category included low birth weight infants, infants of toxemic mothers, infants of diabetic mothers, infants with birth weights over 4,080 g, and twins. On the basis of serial determinations, 7.3% of the risk infants were found to have at least 2 glucose levels of less than 20 mg/100 ml, and 18 had at least 1 level of less than 20 mg/100 ml. Since transient asymptomatic hypoglycemia occurs very frequently in high risk infants, the present regimen of very early oral or intravascular feeding of glucose should be continued in these cases. (15 refs.) - *B. J. Grylack.*

University of Missouri College of Medicine
Columbia, Missouri 65201

1736 **CALLAHAN, EDWARD W., JR.; THALER, M. MICHAEL; KARON, MYRON; BAUER, KARINNE; & *SCHMID, RUDI.** Phototherapy of severe unconjugated hyperbilirubinemia: Formation and removal of labeled bilirubin derivatives. *Pediatrics*, 46(6):841-848, 1970.

Two infants aged 5 and 7 months with severe, congenital, unconjugated hyperbilirubinemia (Crigler-Najjar syndrome) were found to respond to phototherapy with marked and sustained reactions in serum bilirubin levels. During phototherapy, both patients were infused with tracer amounts of ^{14}C-bilirubin for prolonged periods; plasma, urine, and bile were collected; and the disposition of the infused labeled pigment was determined. The study provided conclusive evidence that phototherapy converts a portion of the bilirubin in the body to pigment derivatives that are eliminated rapidly from the blood. This conclusion was based on the fact that radioactive bilirubin derivatives were not detectable in the serum but were plentiful in the bile, particularly during periods of phototherapy. The observations made in this study documented for the first time the finding that light converts a portion of the bilirubin pool in man to more water-soluble derivatives that are excreted efficiently in the bile and urine. (21 refs.) - *B. J. Grylack.*

*University of California Department of Medicine
San Francisco, California 94122

1737 **PLOTKIN, S. A.; & MELLMAN, W. J.** Rubella in the distant past as a possible cause of congenital malformations. *American Journal of Obstetrics and Gynecology*, 108(3):487-489, 1970.

Case studies of 4 congenitally malformed infants conceived 1 year or more after their mothers had been infected with rubella were evaluated, and a survey was done of anomalies in unselected pregnancies following the finding of 1 pregnancy complicated by rubella in order to investigate the possibility of a rubella-carrier state in mothers which has an effect on subsequent pregnancies. The 4 malformed infants were delivered from mothers who had rubella within the 2 years preceding the period of gestation but not during that period. Laboratory studies showed that these malformations were not the result of an intrauterine infection of the malformed fetus with rubella virus. The survey of 86 women who gave birth to infants with congenital rubella syndrome revealed a small but probably insignificant increase in fetal abnormality in subsequent births. Larger epidemiologic studies are necessary before the possibility that maternal rubella is in some cases injurious to more than 1 pregnancy can be excluded. (4 refs.) - *B. J. Grylack.*

Wistar Institute of Anatomy and Biology
Philadelphia, Pennsylvania 19104

1738 **QUEENAN, JOHN T.; GADOW, ENRIQUE C.; BACHNER, PAUL; & KUBARYCH, SANDRA F.** Amniotic fluid proteins in normal and Rh-sensitized pregnancies. *American Journal of Obstetrics and Gynecology*, 108(3):406-414, 1970.

Four hundred and eighty-one amniotic fluid samples from 221 obstetric patients between 12 and 43 weeks of gestation were studied in order to establish base-line values for amniotic fluid proteins in normal and Rh-sensitized pregnancies. Results from normal pregnancies showed in general that the higher the birth weight, the lower the amniotic fluid protein value is prior to delivery. In normal pregnancies and in those with mild and moderate erythroblastosis fetalis, the

protein values appear to decrease as gestation progresses. In the case of severe Rh disease, the protein range is greater and the trend is not always downward with increasing gestation. In the setting of an elevated amniotic fluid bilirubin, a rising protein trend with a final value >0.800 g per 100 ml was found with hydrops fetalis. The study of amniotic fluid proteins may be used as a prognostic tool for evaluation of the severity of erythroblastosis fetalis. Protein values for normal pregnancies may become especially pertinent in view of recent advances in enzyme studies of amniotic fluid for the prenatal diagnosis of genetically related defects. (15 refs.) - *B. J. Grylack.*

New York Hospital-Cornell Medical Center
New York, New York

1739 **STREISSGUTH, ANN PYTKOWICZ; VANDERVEER, BEVERLY B.; & SHEPARD, THOMAS H.** Mental development of children with congenital rubella syndrome. *American Journal of Obstetrics and Gynecology*, 108(3):391-399, 1970.

The mental development of 12 children born with congenital rubella syndrome was followed for 2 years in an attempt to correlate mental development with different variables of the congenital problem. Gestational age at the onset of maternal rubella proved to be a significant variable, with the critical period for maximum impairment falling between 14 and 60 days. Of the 12 children, none could be considered normal at age 2. Although 4 appeared to have normal mental development at that age, the prognosis was guarded because of deafness and microcephaly. No correlation was noted between head circumference and intellectual development. Despite the low predictive validity of an IQ score at 24 months, the degree of sensory handicap and motor and mental delay manifested by these children limits optimistic prognoses. Knowledge of the degree of intellectual and sensory deficit to be expected in a child born with rubella would appear to be valuable information for a woman exposed to rubella during the first trimester of pregnancy. (9 refs.) - *B. J. Grylack.*

University of Washington School of Medicine
Seattle, Washington

1740 **CHESSELLS, JUDITH M.; & WIGGLESWORTH, J. S.** Haemostatic failure in babies with rhesus isoimmunization. *Archives of Disease in Childhood*, 46(245):38-45, 1971.

Disseminated intravascular coagulation was a major cause of hemostatic failure in babies with rhesus isoimmunization. However, impaired hepatic synthesis of coagulation factors should also be considered. Measurements were made of platelet counts, fibrinogen levels, thrombotest, thromboplastin generation screening, thrombin ratio, and titer of circulating fibrin degradation products in 30 infants with Rh isoimmunization. Five infants evidenced a clinical tendency toward bleeding, and there was evidence of disturbed hemostasis in 8. Intravascular fibrin deposits were present in the tissues of 3 out of 4 infants who died of hemorrhage. In an additional retrospective survey, the major factor causing death in 10 of 18 neonates who died with rhesus isoimmunization from 1967 to 1969 was hemorrhage. Necropsy revealed that sites of hemorrhage were mainly intracranial and intrapulmonary. Infants with a cord Hb <7 g/100 ml were at risk from hemostatic failure. (28 refs.) - *J. K. Wyatt.*

Hammersmith Hospital
London, England

1741 **DELANEY, J. J.** Rubella vaccination during the puerperium. *Obstetrics and Gynecology*, 37(4):634-635, 1971.

Of 1,079 women with normal pregnancies who were screened for the presence of rubella hemagglutination-inhibition antibodies, 97 had no detectable antibodies. Fifty-five of these women were available for vaccination after delivery and were vaccinated with the Cendehill strain. Mild symptoms which may or may not have been related to the vaccine were noted in 5 patients. Follow-up of 31 patients over a 6-week period revealed that demonstrable hemagglutination-inhibition antibodies to the rubella virus had failed to develop in 4 (12.9%). A fourfold rise in antibody titer was observed in 18 patients after vaccination. Patients who are vaccinated after pregnancy should have repeat blood studies after vaccination; when immunity fails to develop, they should be revaccinated. (No refs.) - *J. K. Wyatt.*

University of Colorado Medical Center
Denver, Colorado

1742 **KELLY, CHARLES S.; GIBSON, JACK L.; WILLIAMS, CURTIS S.; & LEIBOVITZ, ALBERT.** Postpartum rubella immunization: Results with the HPV-77 strain. *Obstetrics and Gynecology*, 37(3):338-342, 1971.

The immediate puerperium seems to be an ideal time for safe and effective rubella immunization since there is a low risk of inadvertently vaccinating a pregnant woman whose contraception has failed. Two hundred and thirty-six of 1,884 women delivered during a 10 month period lacked antibody to rubella when screened by the hemagglutination-inhibition method. Follow-up data on 201 women who were vaccinated with the HPV-77 strain during the immediate puerperium revealed a 91% incidence of seroconversion, indicating no suppression of the immune response. Side effects occurred in 22.1% of the patients but were transient in all but 2 cases. Incidence of arthralgias was 17.8%. Neither morbidity nor seroconversion were seen in infants of the vaccinees. (20 refs.) - *J. K. Wyatt.*

U. S. Army Hospital
Fort Carson, Colorado 80913

1743 **GUENTER, KENNETH E.; & ESTELA, LUIS A.** Rubella antibody determination among pregnant women in U. S. military communities in Europe. *Obstetrics and Gynecology*, 37(3):343-347, 1971.

The hemagglutination-inhibition (HI) test should be a routine part of the prenatal examination. Of 947 pregnant women who were screened for rubella antibody with the HI test, 98 (10.3%) were susceptible to rubella. Correlation of history of rubella with antibody determination in 874 of these patients disclosed a fairly constant percentage with antibody of 88.4%-91.3% among 804 Caucasians, regardless of history. The correlation for the 64 Negro patients was poorer; 22.5% without antibodies and a positive history, and 8.1% without antibodies and a negative history. Although these findings are based on a small sample, they demonstrate that history of rubella infection is not a reliable guide to immune status. (22 refs.) - *J. K. Wyatt.*

U. S. Army Hospital
Fort Carson, Colorado 80913

1744 **ISACSON, PETER; KEHRER, ANTHONY F.; WILSON, HAZEL; & WILLIAMS, SANDRA.** Comparative study of live, attenuated rubella virus vaccines during the immediate puerperium. *Obstetrics and Gynecology*, 37(3):332-337, 1971.

Of 107 adult women who were given attenuated rubella virus vaccines within 1 week postpartum, 34 received HPV-77 DK_{12}, 38 received HPV-77 DE_5, and 5 received Cendehill. Sixty-six women did not have the rubella HI antibody titer prior to immunization. Titers of 1:10 or 1:20 were present in the remainder. Antibody levels were not significantly different for the 3 vaccines, and seroconversions for each vaccine were comparably high. Duration and the symptoms of limitation-of-motion and swelling were most marked among DK_{12} and least severe among Cendehill recipients. The most common complaint in each group was joint pain (40.9%), and 2 initially seronegative women who had received the DK_{12} strain had joint symptoms which limited their ability to care for their infants. Recurrences were observed in 1 DK_{12} recipient, as long as 8 months after inoculation. A detectable HI antibody titer was not found in 18 infants from 4 to 11 months after the mother had been immunized. Live, attenuated rubella vaccines may be administered safely and effectively to adult females immediately after delivery. (15 refs.) - *J. K. Wyatt.*

2211 Main Street
Buffalo, New York 14214

1745 **DAVIS, LARRY E.; TWEED, GERALD V.; CHIN, TOM D. Y.; & MILLER, GERALD L.** Intrauterine diagnosis of cytomegalovirus infection: Viral recovery from amniocentesis fluid. *American Journal of Obstetrics and Gynecology*, 109(8):1217-1219, 1971.

The presence of intrauterine infection was established during the twenty-first week of gestation by recovery of the cytomegalovirus from a sample of amniotic fluid obtained by amniocentesis. The patient developed anorexia, lethargy, and a low-grade fever at 4 weeks gestation. In addition to these continuing symptoms, she had splenomegaly, thrombocytopenia, lymphocytosis, and

atypical lymphocytosis. Diagnosis was cytomegalovirus mononucleosis. The presence of cytomegalovirus in the amniotic fluid indicated probable infection of the fetus. A therapeutic abortion was performed at 22 weeks of gestation. Autopsy of the fetus revealed the presence of cytomegalovirus infection but no congenital anomalies. Amniocentesis is a valuable method for obtaining material to establish antepartum diagnosis of cytomegalovirus infection. (10 refs.) - *J. K. Wyatt.*

National Communicable Disease Center
Kansas City, Kansas

1746 **VIDYASAGAR, D.** Birthweight in haemolytic disease of newborn. *Archives of Disease in Childhood*, 46(245):113-115, 1971.

Rh hemolytic disease of the newborn (HDN) does not appear to interfere with fetal growth as measured by birthweight. A comparison of the birthweights of 190 neonates with HDN and normal newborns revealed no significant between-group differences after 35 weeks of gestation. At 34 weeks of gestation, weight differences were significant. The birthweights of the 26 infants with HDN who received fetal transfusions were normal, and these infants did not differ from other HDN infants. (8 refs.) - *J. K. Wyatt.*

Children's Hospital
Winnepeg 3, Manitoba, Canada

1747 **ELCHARDUS, J.-F.; LABRUNE, B.; RIBIERRE, M.; & MALLET, R.** Localisation encephalique de la maladie generalisee des inclusions cytomegaliques du nouveau-ne (Brain involvement in cytomegalic inclusion disease of the newborn). *Annales de Pediatrie*, 18(5):390-398, 1971.

Cytomegalic inclusion disease may be contracted *in utero* with alarming frequency (1% of newborns in one series) and with significant sequelae. Neurological symptoms and MR are common. In a study of microcephalics with MR, 10% had cytomegalic inclusions. Symptoms may be ubiquitous and the disease discovered only at autopsy. There is sufficient evidence to recommend a vaccination program against this disease. (39 refs.) - *E. L. Rowan.*

Hopital des Enfants-Malades
F 75 Paris 15^{e}, France

1748 **NAKAO, T.** Prognosis of aseptic meningitis. *Developmental Medicine and Child Neurology*, 12(5):680, 1970. (Letter)

Contrary to the accepted view, the long-term prognosis of aseptic meningitis has not been found to warrant any optimism. The Ss were 618 cases of aseptic meningitis caused by Coxsackie B5 and A9 viruses and contracted during a summer epidemic in northern Honshu, Japan, in 1961. These cases were examined 1 to 8 years after hospital discharge. In 171 of the 618 Ss there were changes, such as inattentiveness, irritability, restlessness, recurrent headache, lowering in scholarship, and occasional convulsions. There was no relationship between the postmeningitic changes and age at onset of illness, causative agent, duration of fever, pleocytosis in CSF, or degree of pleocytosis in CSF. EEGs available in 65 cases revealed abnormal findings in 26 instances. These investigations indicate that the long-term prognosis for neurological, behavioral, and electroencephalographic development is not good, although prognosis for life is good. (2 refs.) - *S. Glinsky.*

Sapporo Medical College
S.1 W.17, Sapporo, Japan

1749 **NISWANDER, KENNETH R.** Spectrophotometric analysis of amniotic fluid in early gestation. *American Journal of Obstetrics and Gynecology*, 108(8):1296-1297, 1970.

Liley zones for predicting severity of intrauterine hemolytic disease cannot be projected backward much farther than 23 to 26 weeks gestation. Spectrophotometric analyses of amniotic fluid were performed on 38 patients undergoing therapeutic abortion at various stages of gestation and on 5 gravida between 21 and 26 weeks gestation who subsequently delivered Rh-negative babies. The total of 46 determinations indicated that pigment content of the amniotic fluid increases from early pregnancy, reaches a maximum level

at about 17 weeks gestation, and remains fairly constant until about week 26, when it begins to drop. The inability to project Liley zones farther backward is balanced by the fact that intrauterine transfusions before this gestational interval are not practical at present. (3 refs.) - *S. Glinsky.*

University of California, Davis
Davis, California

1750 **MARINI, A.; BARBARANI, V.; BONCOMPAGNI, P.; & CATTANEO, F.** Alkalinising agents in the delivery room. *Lancet*, 2(7666):265, 1970.

Tromethamine (THAM) and sodium bicarbonate seem to be equally effective in the correction of acidosis in premature infants under certain circumstances. Although THAM has theoretical and experimental advantages over sodium bicarbonate, a trial was conducted in which THAM and sodium were administered alternately to premature infants with acidosis. The infants all weighed between 1 and 2 kg and had Apgar scores of 5 or under. Initially all received a 15% glucose solution via the umbilical vein within 20 minutes of birth. Later THAM and sodium bicarbonate were given alternately, one-third of the dose being administered in the first 10 minutes and the remainder within 6 hours. The intravenous glucose therapy made little difference in severe respiratory distress (S.R.D.) or total mortality, but there was a sharp decline in both after introduction of early alkali therapy. The mortality rate in S.R.D. patients was only moderately reduced, the figures for both alkalis being exactly the same. No adverse reactions followed either drug, and the rate of acidosis correction was the same for both drugs. It is not yet known whether THAM is more effective than sodium bicarbonate for the rapid correction of acidosis in asphyxiated newborn infants. (9 refs.) - *S. Glinsky.*

University of Milan
Milan, Italy

1751 **PAGET, G. E.** Rubella vaccination. *Lancet*, 2(7666):266, 1970. (Letter)

Women or girls outside of the 11 to 14 year old age group may be vaccinated with the rubella vaccine "Cendevax," provided certain conditions are met. A recent letter from the Department of Health makes available the rubella vaccine "Cendevax" to local health authorities for the mass vaccination of girls between 11 and 14 years of age. Others outside this age group may be vaccinated if they are known not to be pregnant at the time of vaccination and intend not to become pregnant during the next 2 months. The vaccine may be obtained through normal pharmaceutical channels. (No refs.) - *S. Glinsky.*

Smith Kline and French Laboratories, Ltd.
Welwyn Garden City, Herts., England

1752 **SEEDS, A. ELMORE; SCHRUEFER, JOHN J. P.; BISSONNETTE, JOHN M.; LIM, HENRY S.; HELLEGERS, ANDRE E.; & BRUNS, PAUL D.** Changes in fetal acid-base chemistries following tromethamine (THAM) or bicarbonate infusions into fetal circulation or amniotic fluid. *American Journal of Obstetrics and Gynecology*, 108(8):1245-1250, 1970.

Direct infusion of bicarbonate or tromethamine (THAM) into the fetus produces equivalent rises in fetal pH, but infusion of THAM into amniotic fluid leads to progressive fetal acidosis. The Ss were 8 rhesus monkeys between 155 and 168 days of gestation, of which 4 received direct infusions of bicarbonate or THAM into the fetus, and 4 received intra-amniotic infusion of THAM. Direct fetal infusion resulted in roughly equivalent increases in fetal pH, but intra-amniotic infusion was not followed by fetal alkaline changes but by progressive fetal acidosis and deterioration. This sharply contrasts with previous experimental success in elevating fetal pH via amniotic fluid bicarbonate infusions. (6 refs.) - *S. Glinsky.*

Johns Hopkins School of Medicine
Baltimore, Maryland

1753 **LARSON, H. ELLIOTT; PARKMAN, PAUL D.; DAVIS, WILLIAM J.; HOPPS, HOPE E.; & MEYER, HARRY M., JR.** Inadvertent rubella virus vaccination during pregnancy. *New England Journal of Medicine*, 284(16):870-873, 1971.

That chronic placental infection can be produced in pregnant women by live rubella virus vaccine is demonstrated by an analysis of 9 cases in which pregnant women were inadvertently so inoculated. One of the women experienced seroconversion; she and another subject exhibited symptoms consistent with vaccine-virus infection. Virus was recovered from the aborted fetuses of both of these subjects; tissues obtained from the remaining 7 subjects yielded no virus. Although viral strains in the placenta do not necessarily establish a teratogenic potential, physicians should be cautious in recommending rubella vaccine to women of childbearing age, since inadvertently immunized pregnant women generally resort to therapeutic abortion, which produces the very fetal loss the vaccine was designed to prevent. (18 refs.) - *N. Jarvis.*

National Institutes of Health
Bethesda, Maryland 20014

1754 Allergies may be at the root of some neurological problems. *Expectations,* 1(1):1, 1971.

Behavioral and mental symptoms that often lead to diagnoses of retardation or emotional disturbance may actually be caused by allergies to food and substances in air and water. In some cases, such symptoms have disappeared after these allergens have been removed from patients' diets or environments. A film presented by Dr. Marshall Mandell at a meeting of the American College of Allergists demonstrated that subcutaneous injection of food substances and synthetic ethyl alcohol produced behaviors that could be mistaken as symptoms of instability or retardation. Mental and behavioral symptoms of this type are described as cerebral allergies. Thus the brain should be regarded as a complex allergic shock tissue that is especially vulnerable because of its excellent blood supply. A center for treatment and study of such ecologic mental illness is being established in Norwalk, Connecticut. (No refs.) - *M. H. Street.*

MEDICAL ASPECTS — Etiologic Groupings

Trauma or physical agents

1755 **MURDOCK, A. I.; LINSAO, L.; REID, M. McC.; SUTTON, M. D.; TILAK, K. S.; ULAN, O. A.; & *SWYER, P. R.** Mechanical ventilation in the respiratory distress syndrome: A controlled trial. *Archives of Disease in Childhood,* 45(243):624-633, 1970.

Artificial mechanical respirators are useful in the management of infants with respiratory distress syndrome (RDS), and the success of the ventilation improves with increased birthweight. In a controlled study of mechanical ventilation (utilizing 3 types of respirators) of infants with RDS, 168 patients received ventilation and 53 did not. Among infants weighing more than 2000g, survival rates improved from 15% (4 of 27 not ventilated) to 43% (29 of 67 ventilated). Ventilation of infants weighing more than 1500g and developing RDS at less than 38 hours of age was associated with improved survival rates (16 of 31) compared to infants of the same weight but ventilated after 38 hours of age (24 of 78). Ventilation of infants weighing 1001 - 1500g was associated with occasional survival; however, none of the unventilated babies in this weight range survived. Among infants weighing more than 2000g the degree of improvement in $[H^+]_2$ was correlated with eventual survival in that fall of $[H^+]_2$ was significantly greater among survivors. Ventilation was associated with improvements within 1 hour in blood gas and in acid-base status in all weight categories. Comparable survival rates were obtained for all 3 respirators tested. (28 refs.) - *M. S. Fish.*

*The Hospital for Sick Children
Toronto 2, Ontario, Canada

1756 **MORDOCK, JOHN B.** Behavioral problems of the child with minimal cerebral dysfunction. *Physical Therapy* 51(4):398-404, 1971.

A review of research data on 5 behaviors frequently cited as characteristic of brain-impaired

children considered hyperactivity, emotional lability, disorders of attention, impulsivity, and general coordination defects. Emotional lability, disorders of attention, and impulsive behavior did not occur more frequently in brain-impaired children than in other emotionally disturbed children. Hyperactive behavior was characteristic of a specific subgroup of brain-impaired children. Behaviors which were found in the majority of brain-impaired youth included physical inadequacy, social inadequacy, and coordination defects. It may be that behaviors originally considered characteristic of the brain-impaired are responses to frustration and anxiety caused by failure. Remedial approaches should be designed to modify the total behavioral difficulty and may require treatment of the child and family. (31 refs.) - *J. K. Wyatt.*

Astor Home and Child Guidance Centers
Rhinebeck, New York 12572

1757 **REYNOLDS, E.O.R.** Indications for mechanical ventilation in infants with hyaline membrane disease. *Pediatrics,* 46(2):193-202, 1970.

During a 3-year period, 104 infants with hyaline membrane disease who weighed more than 1,000g at birth were treated at a hospital center. The question of need for mechanical ventilation was an important aspect of the decision-making process in most cases. Seventy-two infants survived without mechanical ventilation. The other 32 infants became apneic or collapsed with a heart rate of less than 80 per minute and were ventilated mechanically after simple resuscitative measures failed. Seven of these 32 infants survived. The prognosis for mechanically ventilated infants was extremely poor if they had to be ventilated within 24 hours of birth. If apnea does not appear until after 24 hours, however, mechanical ventilation has a substantial hope of success. This treatment should be avoided if possible, due to its attendant risks of infection, interruption of normal pulmonary secretion drainage mechanisms, and possible association with pulmonary fibrosis. (26 refs.) - *B. J. Grylack.*

University College Hospital Medical School
London, W.C. 1, England

1758 **BRUMLEY, GEORGE W.** The critically ill child: The respiratory distress syndrome of the newborn. *Pediatrics,* 47(4):758-769, 1971.

With a proper treatment regimen, infants with respiratory distress syndrome (RDS) due to hyaline membrane disease are able not only to survive but also to have the potential for normal development. Blood gas and pH studies and chest X-rays are essential for confirmation of the diagnosis and the monitoring of therapy. Temperature support to achieve a neutral thermal environment, sodium bicarbonate infusion to correct acidosis, oxygen administration to maintain circulation, and fluid and glucose infusion feedings are essential. Antibiotics and phototherapy are also used routinely. Ventilatory assistance may be necessary in more severe cases. Maximal benefit is obtained in a well-staffed and carefully monitored intensive care nursery. (43 refs.) - *E. L. Rowan.*

Duke University Medical Center
Durham, North Carolina 27706

1759 **NAEYE, RICHARD L.; HARCKE, HOWARD T.; & BLANC, WILLIAM A.** Adrenal gland structure and the development of hyaline membrane disease. *Pediatrics,* 47(4):650-657, 1971.

Adrenal glands were found to be 19% lighter in a sample of 222 infants with hyaline membrane disease than in a control sample of 165 neonates who died of other causes. Control infants had more cells in both permanent and fetal cortices. Adrenal glands were heavier in infants with infections both with and without hyaline membranes and the number of infants with both findings was relatively small. A group of 14 anencephalic neonates with remarkably small adrenal cortices also showed a 55% decrease in the mass of osmiophilic granules in pulmonary type II alveolar cells (the anatomic representation of surfactant). The temptation to treat all infants at risk for hyaline membrane disease with steroids should be resisted until the mechanisms in question have been further explored. (21 refs.) - *E. L. Rowan.*

M. S. Hershey Medical Center
Hershey, Pennsylvania 17033

1760 **REYNOLDS, JOHN W.** The adrenal cortex, steroids, and hyaline membrane disease. *Pediatrics,* 47(4):645-646, 1971.

Despite evidence correlating a small adrenal cortex and decreased corticosteroids with hyaline membrane disease, further studies must be undertaken before steroids become the treatment of choice in this condition. Glucocorticoids may stimulate surfactant activity in sheep and rabbits, but animal physiology is not human physiology, and supra-physiologic doses of drugs may have other effects on other fetal organ systems. Maternal cortisol may be found in the fetal circulation despite fetal adrenal cortical insufficiency. Estrogen, whose precursor is also produced by the adrenal cortex, is also diminished in hyaline membrane disease and the decrease in either or both of these products may be secondary to another, as yet unknown, factor. More carefully controlled studies are needed before any treatment modality is recommended for the respiratory distress syndrome. (11 refs.) - *E. L. Rowan.*

311 Pleasant Avenue
St. Paul, Minnesota 55102

1761 **MATOTH, YEHUDA; ZAIZOV, RINA; & FRANKEL, JACOB J.** Minimal cerebral dysfunction in children with chronic thrombocytopenia. *Pediatrics*, 47(4):698-706, 1971.

Among 20 patients followed for chronic idiopathic thrombocytopenia purpura (ITP) were 6 who had come to attention because of serious behavior problems and learning disabilities. These 6 showed visual-motor deficiencies on the Bender-Gestalt test, diminished abstracting ability, and EEG abnormalities consistent with a diagnosis of minimal brain dysfunction (MBD). Only 3 of the ITP children showed no evidence suggestive of MBD. Seven had EEG abnormalities. The prognosis for children with ITP might be reconsidered in view of this evidence of organic brain disease. (13 refs.) - *E. L. Rowan.*

Beilinson Hospital
Petah-Tikva, Israel

1762 **KENNY, THOMAS J.; & CLEMMENS, RAYMOND L.** Medical and psychological correlates in children with learning disabilities. *Journal of Pediatrics*, 78(2):273-277, 1971.

Extensive medical evaluation of children with learning disabilities is relatively unfruitful. The Ss were 100 randomly selected children with learning and/or behavioral problems who were referred to a diagnostic and evaluation clinic because of suspected minimal brain dysfunction. Twenty children each were selected at the age levels 8 to 12 years; the group included 85 males and 15 females, 57 Caucasians, and 43 Negroes. After each child received a thorough medical and psychological examination, electroencephalograms were done on 88 of the children. A *chi* square interaction analysis was set up to compare the results of the neurologic examination, electroencephalogram, and final diagnosis. There was no significant relationship among any of these factors, nor was there any combined or additive effect. The results indicate that the final diagnosis is more contingent upon symptomatology and psychological findings than upon any specific medical, neurologic, or electroencephalographic findings. Considering the poor correlation of the neurologic examination and the electroencephalogram with the final diagnosis, there appears to be minimal utility in referring such children routinely for these procedures. (12 refs.) - *S. Glinsky.*

University of Maryland Medical School
Baltimore, Maryland

1763 **RICHARDSON, ALAN; HIDE, T.A.H.; & EVERSDEN, I. D.** Long-term continuous intracranial-pressure monitoring by means of a modified subdural pressure transducer. *Lancet*, 2(7675):687-690, 1970.

The successful use of a small subdural pressure transducer to monitor intracranial pressure continuously is reported. The technique was particularly helpful in 3 patients with head injuries, but it was of minimal value in 11 patients with intracranial tumors. Pressure-induced cerebral ischemic attacks with no clinical warning, possibly triggered by anesthesia, may be monitored by this method. (18 refs.) - *E. Kravitz.*

St. George's Hospital at
Atkinson Morley's Hospital
Wimbledon, London S.W.20, England

1764 **KEMPE, C. HENRY.** Paediatric implications of the battered baby syndrome. *Archives of Disease in Childhood*, 46(245):28-37, 1971.

Although the true incidence of the battered baby syndrome is not known, it may be as high as 6/1,000 live births. In a group of over 400 battering parents, all social classes, races, creeds, religions, and levels of education and income were proportionally represented. About 5% of battering families have 1 parent with a delusional or depressed type psychosis or who has an encapsulated psychosis regarding a specific child. Another 5% seem to be aggressive psychopaths, and the remaining 90% have serious problems in mothering. Among the clinical findings of the extended battered child syndrome are failure-to-thrive due to deficient mothering, subtle physical findings, and mothers with unjustified complaints about a child. For battering to occur, there must be potential within the parent for abuse, the parents must have a distorted view of the child, and a crisis must occur. Physicians must be willing to make a diagnosis of battering and to admit the child to the hospital in suspected cases. In 80% of the cases where the problem surrounds maternal deprivation of parents, a therapeutic person can become involved in providing mothering therapy and the child can be gradually returned to the home. Parental rights are permanently terminated in 5% of the cases. A reporting system should be developed for cases of suspected parent-inflicted injury and failure-to-thrive due to deprivation. Since these injuries are rarely premeditated or purposely inflicted, reporting systems should not be based on the question of "willful infliction." (2-item bibliog.) - *J. K. Wyatt.*

University of Colorado Medical Center
Denver, Colorado

1765 **TOUWEN, B.C.L.** Neurological follow-up of infants born after obstetrical complications. In: Stoelinga, G.B.A.; & Van Der Werff Ten Bosch, J. J., eds. *Normal and Abnormal Development of Brain and Behaviour.* Baltimore, The Williams & Wilkins Co., 1971, p. 179-186.

A blind follow-up study of 167 children, ages 4 years, 9 months to 5 years, 2 months, all with a large variety of prenatal and perinatal complications, showed that 26% resulted from a pregnancy and delivery without, or with at most one, nonoptimal condition; 66% experienced 2 to 6 nonoptimal obstetrical conditions; and only 8% experienced 7 or more such conditions (these children were not necessarily brain-damaged). In respect to neonatal neurologic data, 52% had 5 or fewer deviations, and the remainder had 6 or more. (Optimal is not the same as classical normality—it merely helps to identify a defined group within the normal range.) Many of the nonoptimal babies showed various neurologic symptoms, but only 2 children showed a major neural dysfunction. A positive relationship was found between neurologic results and sex. An optimal score demonstrated a clear-cut differentiation within a group of so-called clinically "normal" children. (4 refs.) - *B. Berman.*

1766 **DELACATO, CARL H.** Brain injuries and poor reading. In: Delacato, C., *The Treatment and Prevention of Reading Problems.* Springfield, Illinois, C. C. Thomas, 1971, p. 45-53.

Research has shown that boys have reading problems 4 times more frequently than girls; that boys, at birth, have larger heads than girls; and that a severe reading problem may result from anoxia and the consequent brain damage during or subsequent to birth. (Studies of intrauterine oxygen level during birth are seeking to prevent any degree of anoxia.) Clinical symptoms relative to spoken language of more than 200 brain-injured aphasic children with concomitant motor dysfunction differed only in degree from those of the retarded reader relative to written language. In cases of purely traumatic etiology, the medical approaches are examined. (2 refs.) - *B. Berman.*

1767 **BRINK, JOYCE D.; GARRETT, ALICE L.; HALE, WILLIAM R.; WOO-SAM, JAMES; & NICKEL, VERNON L.** Recovery of motor and intellectual function in children sustaining severe head injuries. *Developmental Medicine and Child Neurology,* 12(5):565-571, 1970.

Children who had sustained severe head injuries showed significant motor function recovery but severe deficits in intellectual abilities. The Ss were 52 patients between 2 and 18 years of age, of whom 39 were males and 13 females. All but 10 were injured in motor vehicle accidents. After initial medical attention an active rehabilitation

program of physical therapy, occupational therapy, pediatric medical supervision, and orthopedic care was carried out in each case. Significant recovery of motor function was observed in a majority of the cases, independence in ambulation and self-care being achieved in 87% of the Ss. But severe deficits were observed in the intellectual level measured by intelligence test scores. Younger age group children showed significantly greater impairment than the adolescents. The eventual degree of recovery of both motor function and intellectual ability was related to the duration of coma. In some cases, improvement in both physical and intellectual performance continued for as long as 3 years after injury. (8 refs.) - *S. Glinsky.*

Rancho Los Amigos Hospital
Downey, California 90242

1768 **GARDNER, RILEY W.** Evolution and brain injury: The impact of deprivation on cognitive-affective structures. *Bulletin of the Menninger Clinic*, 35(2):113-124, 1971.

A refinement of Darwinian theory holds that the basic dictate of evolution is: "Survive—by maintaining progressive equilibrations within adaptively tolerable limits—to maximize your genetic potential." The formation of an ever-more-refined hierarchy of cognitive-affective structures is the means by which the developing human organism moves from primitive infantile reactions to subtly differentiated adult equilibrations in a complex social world. Brain injury interferes not only with the unfolding of the adaptive cognitive structure formations but also with the equilibration of the total organism. The human infant unfolds his genetic potential by perceiving parental models and assuming them into his own personality organization. A major, but generally ignored, effect of brain injury or other true deprivation is a massive constriction of self-esteem which is an essential determinant of the less-than-optimal clarity, reality, and subtlety of cognition of the self, parent, and other persons. The truly deprived person is lacking in self-esteem in a rather total sense, which is not the same as the common statement that his self-esteem is low. Yet, the brain injured child valiantly struggles against these overwhelming odds to maximize his genetic potential. (15 refs.) - *J. C. Moody.*

Menninger Foundation
Topeka, Kansas 66601

1769 **DYMENT, PAUL G.; LATTIN, JOHN E.; & HEBERTSON, LEON M.** The value of the electroencephalogram in evaluating children with minimal cerebral dysfunction. *Journal of School Health*, 41(1):9-11, 1971.

Review of current medical opinion concerning the value of the EEG in evaluating children with the minimal cerebral dysfunction syndrome (MCD) suggests that an EEG is not necessary. Although EEG abnormalities in the form of mild diffuse or occasional focal dysrhythmias are frequently found in children with MCD, the EEG is commonly reported as "mildly and diffusely abnormal," "borderline," or "compatible with a seizure disorder," which is no help in determining further diagnostic study, referral, or treatment. Furthermore, various studies have found abnormal EEG tracings in approximately 10 to 30% of normal children. One study reported normal EEGs in 53 out of 106 children (50%) with MCD. Unfortunately, the performance of an EEG is often considered the mark of a thorough evaluation by parents or referral sources. While it may occasionally be wise to have an EEG for everyone's peace of mind if the referral source or the parents are adamant, school placement committees should be discouraged from requiring it for admission to special education classes. (10 refs.) - *J. C. Moody.*

William Beaumont General Hospital (U.S. Army)
El Paso, Texas 79920

MEDICAL ASPECTS – Etiologic Groupings

Disease or disorders of metabolism, growth, or nutrition

1770 **HERNDON, ROBERT M.; RUBINSTEIN, LUCIEN J.; FREEMAN, JOHN M.; & MATHIESON, GORDON.** Light and electron microscopic observations on Rosenthal fibers in Alexander's disease and in multiple sclerosis. *Journal of Neuropathology and Experimental Neurology*, 29(4):524-551, 1970.

Five cases are reported in which Rosenthal fibers, an accumulation of rod-shaped eosinophilic material within astrocytes, were found. Three of these cases are attributed to the presence of Alexander's disease, characterized by megancephaly and progressive spasticity and dementia beginning in infancy, and 2 show histologic evidence of multiple sclerosis. In Alexander's disease the Rosenthal fiber formation is generalized throughout the brain and spinal cord, but in multiple sclerosis and in other diseases it appears only in altered astrocytes and in reactive astrocytes in and around demyelinated areas. It is hypothesized that an astroglial enzyme defect becomes manifest at the time of myelination in the infantile cases of Alexander's disease. In cases of Alexander's disease of late onset and in the cases of multiple sclerosis, the metabolic abnormalities were latent, but became manifest in the presence of some other central nervous system disease. (33 refs.) - *E. L. Rowan.*

Johns Hopkins University School of Medicine
Baltimore, Maryland 21205

1771 **LAKE, BRIAN D.; STEPHENS, ROSEMARY; & NEVILLE, B.G.R.** Syndrome of the sea-blue histiocyte. *Lancet*, 2(7667):309, 1970. (Letter)

A comparison of reported descriptions of the syndrome of the sea-blue histiocyte with studies of 4 Ss having these cells in the marrow suggests the possibility that 2 conditions in which such cells are present may exist. Follow-up of the 4 patients, 2 sibs presenting with splenomegaly in the first year of life and 2 additional patients, has shown the disease in these 4 Ss to be a progressive one; a neurological disorder with ataxia, dementia, and seizures developed in the sibs in later years. The other 2 patients also had a progressive neurological disease. These findings are in contrast to the previously reported benign course of the disease. Follow-up of the benign patients would aid in the resolution of the discrepancy or indicate if 2 separate conditions exist. The use of the term sea-blue histiocyte is also questionable, since the marrow of these patients contains another variety of cells which do not stain this color. (5 refs.) - *M. S. Fish.*

Hospital for Sick Children
London W.C.1, England

1772 **LESCH, MICHAEL.** The purine revolution. *New England Journal of Medicine*, 283(22):1222, 1970. (Editorial)

The years 1963-1970 can be termed the "purine revolution" since during that period there were many thorough revisions of theories on the metabolic control of purine biosynthesis and the classification and treatment of hyperuricemia. Development of a technic for determining *in utero* whether the male fetus of a mother who is heterozygous for hypoxanthine-guanine phosphoribosyl transferase (HGPRT) activity had inherited the mutant X-linked gene and would thus exhibit the syndrome of choreathetosis, mental retardation, hyperuricemia and self-mutilation is an example of altered thinking on purine metabolism. This is made possible by assays of fibroblast cultures obtained from amniotic-fluid fibroblasts for sex chromatin and HGPRT activity. Therapeutic abortion is recommended when total HGPRT deficiency is demonstrated. (9 refs.) - *G. S. Bell.*

No address

1773 **GOMES, W. J.; & *HUNTER, JANET L. P.** Mental retardation, cataracts, and unexplained hyperphosphatasia. *Archives of*

Disease in Childhood, 45(243):726-727, 1970.

Examination of a 4-year-old male disclosed that the S was MR, had cataracts, and had a sharply raised level of serum alkaline phosphatase, 90% of which was attributable to bone isoenzyme, despite the lack of evidence of a bone disease. His mother's pregnancy was mildly toxemic. After a cyanotic attack at 4 days postnatal, the child's first year was uneventful except for delayed milestones. At 20 months a convulsion occurred, followed by 2 additional ones during the third year despite anticonvulsant therapy. Increase in the dose level of the drug lessened the severity of the convulsions. Psychological examination at 4 years 4 months showed a low normal level of intelligence. Electroencephalographic recordings showed gross abnormalities, suggestive of grand mal epilepsy. Clinical findings were normal, except for vision, which was poor due to cataracts. Laboratory findings of blood and urine specimens were normal except for elevated alkaline phosphatase levels, which were 376, 88, and 77 KA units/ml, respectively, on 3 separate samples. Isoenzyme electrophoresis of 1 of the samples gave a band at Rf 0.56. Destruction of 90% of the material after incubation at 56°C for 15 minutes indicated osseous origin. A full skeletal survey by X-rays showed no radiological abnormality. (3 refs.) - *M. S. Fish.*

*Scartho Road Hospital
Grimsby, England

1774 STIMMLER, L.; JENSEN, N.; & TOSELAND, P. Alaninuria, associated with microcephaly, dwarfism, enamel hypoplasia, and diabetes mellitus in two sisters. *Archives of Disease in Childhood*, 45(243):682-685, 1970.

A study of 2 sisters, born 1½ years apart, has disclosed severe MR associated with microcephaly, dwarfism, enamel hypoplasia, and elevated levels of urinary alanine. Both physical and mental development remained retarded. Developmental level of 1 sib was 18 months at CA of 5 years 5 months. The corresponding level for the younger sib was 8 months at CA of 4 years. The sisters became diabetic, but insulin stabilized this latter disorder. Chromatography of urinary amino acids disclosed abnormally high alanine levels; plasma alanine levels, both fasting and after glucose loads of 1g/kg, were well above the range for normal and diabetic controls. Blood pyruvate and lactate fasting levels were also above the range for the 2 control groups; these levels rose 3- to 4-fold after the glucose loads; whereas they rose no more than 60% in the normal control and diabetic groups. While 2 other syndromes associated with raised lactate and pyruvate levels are known (one of them also associated with MR), little other clinical correlation with these 2 patients exists, indicating that underlying defects are probably different for each group. (11 refs.) - *M. S. Fish.*

Guy's Hospital
London S.E. 1, England

1775 VAN SANDE, MARC. Hair amino acids: Normal values and results in metabolic errors. *Archives of Disease in Childhood*, 45(243):678-681, 1970.

An examination of the amino acid content of hair has indicated that a biochemical relationship may exist between inborn errors of amino acid metabolism and morphological hair abnormalities. Quantitative determination of 17 separate amino acid levels, utilizing a Technicon Amino Acid Analyzer, from hydrolyzed hair samples obtained from 25 patients suffering from metabolic errors and 10 normal controls showed that hair phenylalanine levels were significantly higher and glutamic acid levels were lower in samples from 7 patients with phenylketonuria (PKU) than in those from normal Ss. Phenylalanine levels were also increased in samples from 5 patients with homocystinuria. Cystine levels for samples from this latter group were normal; however, pyridoxine therapy for this group may have contributed to this finding. Comparison of observations in this study with those reported by other investigators is difficult since insufficient control samples were analyzed in previous studies. The finding that for disorders such as argininemia and histidinemia high concentrations of 1 amino acid are found in biological fluids but are not present in hair, whereas for PKU and glycinemia a high concentration occurs both in the fluids and in hair, merits further inquiry. (12 refs.) - *M. S. Fish.*

Born-Bunge Stichting
F. Williotstraat, 2600-Berchem, Belgium

1776 **BENTOVIM, A.; *CLAYTON, BARBARA E.; FRANCIS, DOROTHY E. M.; SHEPHERD, JEAN; & WOLFF, O. H.** Use of an amino acid mixture in treatment of phenylketonuria. *Archives of Disease in Childhood,* 45(243):640-650, 1970.

An initial evaluation of the dietary use of an amino acid mixture for phenylketonuric (PKU) children instead of a commercial preparation of a low phenylalanine protein hydrolysate has indicated good acceptance of the diet and reduction in food and family problems. Of 12 PKU children (ages from 21 months to 6 years 2 months) who had been diagnosed and treated early in life with a commercial preparation (Cymogran) substituting for natural protein, 6 received an amino acid mixture (3 DL amino acids and the remainder L, only) for 6 months, along with cereal, milk, potatoes, fruit, vegetables, low phenylalanine bread, and appropriate quantities of fat, carbohydrates, minerals, and vitamins. The other 6, paired by age with the experimental group, continued on the commercial diet. Laboratory findings and psychological evaluations of the experimental group showed that normal growth was maintained and that plasma and urinary amino acid levels were normal. Overall food problems, particularly refusal, vomiting, and dawdling, were reduced in the experimental group, and family tensions associated with feeding lessened. The emotional state of 2 of the 3 disturbed children in this group improved. While the amino acid mixture appears to be of value for this particular age group, any recommendations for its use by younger PKU children should be preceded by further evaluation. (23 refs.) - *M. S. Fish.*

*The Hospital for Sick Children
London W.C.1, England

1777 **MAYO, O.; & SUTCLIFFE, R. G.** Is lyonisation total in man? *Lancet,* 2(7671):528, 1970. (Letter)

A hypothesis that an enzymatic process outside of the red-cell membrane determines Xg antigenic specificity may explain an observation that erythrocytes from women heterozygous at the Xg locus cannot be separated into Xg(a+) and Xg(a-). The hypothesis thus suggests that the mode of construction of the Xg antigen, and not the extent of X-inactivation, is important in this process and provides a more simple (and testable) explanation than the one based on the assumption that each erythrocyte has its Xg specificity determined solely by its own nuclear genes. The best evidence for X-inactivation in man is that individual clones of cultured fibroblasts from females heterozygous for the glucose-6-phosphate dehydrogenase A+B+ polymorphism can be shown to express only one or the other of the alleles at the locus. Similar observations have been made on fibroblasts cultured from carriers of Lesch-Nyhan syndrome, where the deficient enzyme is hypoxanthineguanine phosphoribosyltransferase. Evidence that other antigens may be produced extracellularly also supports this hypothesis. A simple test of the hypothesis would be the attempted conversion of Xg(a-) cells to Xg(a+) cells by incubation with extracts of Xg(a+) marrow tissue from homozygous or hemizygous Xg(a+) individuals. (13 refs.) - *M. S. Fish.*

Western General Hospital
Edinburgh, EH4 2HU, Scotland

1778 **KELLY, SALLY; KORNS, ROBERT; & BURNS, JEAN.** Sex ratio in phenylketonuria. *Lancet,* 2(7667):314-315, 1970. (Letter)

A screen for phenylketonuria (PKU) of over 1.5 million infants born in New York State during 1965-1969 has not reflected the highly altered sex ratios reported in another study. The sex ratio in the present study was 1.24 in favor of males (67 males and 55 females with PKU) and compares favorably with values observed in a Massachusetts study where the sex ratio for PKU was 1.20 (24 males and 20 females). Comparison of reported birth weights of the PKU infants with values for normal babies indicated that the greater frequency of abnormal phenylalanine levels in males was not related to prematurity, although lower birth weights may be a factor affecting phenylalanine levels. (5 refs.) - *M. S. Fish.*

Birth Defects Institute
New York State Department of Health
Albany, New York

1779 **SAUGSTAD, LETTEN F.** Sex ratio in phenylketonuria. *Lancet,* 2(7667):314, 1970. (Letter)

Analysis of a screening program for phenylketonuria (PKU) among newborns has shown that of the entire group of 81 cases of PKU, only one-third had a normal birth history, and these latter Ss included twice as many females as males, a finding in agreement with other observations that increased perinatal morbidity occurs among male infants. This morbidity may contribute to increased phenylalanine levels such as those found associated with jaundice. These findings suggest the possibility that a previously reported study may have missed PKU babies with a normal birth history and, for this reason, resulted in the diagnosis for PKU of twice as many males as females. The present study may also have identified a high proportion of males with PKU had it been limited to screening babies at risk. (8 refs.) - *M. S. Fish.*

E.E.G. Laboratory
KyrreGrepps-gt. 11
Oslo 4, Norway

1780 **FIALKOW, PHILIP J.** Is lyonisation total in man? *Lancet*, 2(7667):315, 1970. (Letter)

The Lesch-Nyhan syndrome is an inherited X-linked recessive trait characterized by hyperuricemia, MR, choreoathetosis, and compulsive self-mutilation. Findings in heterozygous females support an explanation of recent observations that inactivation at the Xg locus was not detected in patients with myelocytic leukemia. Red cells and skin fibroblasts of Ss with Lesch-Nyhan syndrome have almost no hypoxanthine-guanine phosphoribosyltransferase; however, skin fibroblasts of heterozygous females have an intermediate level of the enzyme activity, and their red cells have normal activity. If red cells of heterozygotes are derived from X chromosomes with the normal gene for the enzyme, while an inactivated X chromosome bears the allelic Xg^a gene, red cells should be Xg(a-) according to the Lyon hypothesis; however, in all 3 reported cases where the Xg gene was on the same chromosome as the gene for normal enzyme activity, an Xg(a+) phenotype was observed. If the Xg^a antigen is synthesized in red cells, these observations support previous suggestions that the Xg locus does not undergo random fixed inactivation when present on a normal chromosome. (4 refs.) - *M. S. Fish.*

University of Washington
Seattle, Washington

1781 **WIESMANN, ULRICH; & NEUFELD, ELIZABETH F.** Metabolism of sulfated mucopolysaccharide in cultured fibroblasts from cystic fibrosis patients. *Journal of Pediatrics*, 77(4):685-690, 1970.

Examination of the turnover of mucopolysaccharide in fibroblasts from 13 patients with cystic fibrosis showed that the sulfated mucopolysaccharide metabolism differed only slightly, if at all, from normal and could easily be differentiated from that of the mucopolysaccharide storage disorders—Hunter's and Hurler's syndromes. Intracellular accumulation of radioactive mucopolysaccharides in fibroblasts from cystic fibrosis patients over a 3-day period ranged from 0.8 to 1.9 times the normal level. Six patients with Hurler's syndrome and 7 patients with the Hunter syndrome evidenced accumulations from 3.5 to 9 times the normal load over the same period. However, fibroblasts from one Hurler line consistently accumulated only 2 to 3 times as much as normal during a 3-day period, and the accumulation pattern could not be readily differentiated from the fibroblasts of cystic fibrosis patients at the high end of the accumulation range. A pulse-chase experiment readily distinguished between the Hurler and cystic fibrosis conditions. The cystic fibrosis gene appears to cause some undefined defect which affects a number of metabolic pathways, including those of the mucopolysaccharide turnover. (12 refs.) - *J. K. Wyatt.*

National Institutes of Health
Bethesda, Maryland 20014

1782 **HALL, W. KNOWLTON; CRAVEY, C. EUGENE; CHEN, PING TUAN; OSTENDORFF, MARTHA E.; HOLLOWELL, JOSEPH G.; & THEVAOS, THEO G.** An evaluation of galactosuria. *Journal of Pediatrics*, 77(4):625-630, 1970.

Tests for urinary carbohydrate excretion performed on 1,200 children, most of whom were institutionalized MRs, identified 59 children who regularly excreted lactose in the urine following breakfast. Results were variable because of the variation in carbohydrate intake by the children. A more definitive screening procedure was obtained when tests were performed after a controlled, oral galactose load than when testing was done on random postprandial urine samples. The use of this screening procedure and blood sugar and galactose-1-phosphateuridyl transferase assays

led to the identification of 2 children with galactosemia and others with significant renal defects. (16 refs.) - *J. K. Wyatt.*

Medical College of Georgia
Augusta, Georgia 30902

1783 **LOENING, W.E.K.; & VINIK, A. I.** The association of growth hormone deficiency and pituitary myxoedema. *Helvetica Paediatrica Acta*, 25(2):114-118, 1970.

A combination of human growth hormone (HGH) deficiency and signs of pituitary myxedema was observed in a 12-year-old African female. The girl was brought to medical attention because of growth failure, lethargy, and lack of interest in her environment. Radiological examination showed femoral epiphyseal dysgenesis; laboratory tests revealed evidence of human growth hormone and thyroid stimulating hormone (TSH) deficiencies. Since the family of the girl moved away, significant follow-up on mental and physical status is lacking. The etiology of anterior pituitary hormone deficiencies, other than neoplastic, remains obscure. Treatment of thyroid stimulating hormone deficiency consists of thyroxine administration on a longterm basis. Replacement therapy with human growth hormone is more difficult. Administration of testosterone would result primarily in skeletal maturation and a growth spurt. (21 refs.) - *B. J. Grylack.*

University of Natal
Durban, Natal, South Africa

1784 **FONTAINE, G.; FARRIAUX, J. P.; & DAUTREVAUX, M.** L'hyperprolinemie de type I. Etude d'une observation familiale (Type I hyperprolinemia. Study of a family). *Helvetica Paediatrica Acta*, 25(2):165-175, 1970.

In a 3-month-old female infant with familial hyperprolinemia type I, systematic description of the aminoacidopathies led to the detection of a hyperprolinemia which confirmed the specific levels of amino acids in the blood and urine. This type of hyperprolinemia, characterized by the absence of Δ_1-pyrroline-5-carboxylic (PC) acid, is asymptomatic clinically and biologically. Two tests were made on the S with loading of 1-proline. In the blood, an important peak of prolinemia was noted with persistence of a hyperprolinemia until the twenty fourth hour. In the urine, there was no significant modification of the diagram of amino acids. Study of the parents and 7 sisters of the girl by oral loading with 1-proline manifested levels of abnormal prolinemia, with abnormalities designated in the curve of Ss who were also unaffected clinically. A normal level of proline was retained in 3 sisters, while the level was elevated moderately in the mother and was clearly elevated in the father and 4 sisters. (22 refs.) - *B. J. Grylack.*

Service de Pediatrie de la Cite Hospitaliere
Lille, France

1785 **MARTINOV, Y. S.; MALKOVA, E. V.; & KOUZNETSOV, V. N.** Troubles neuropsychiques dans les affections du foie, des voies biliaires et du pancreas (Neuropsychiatric difficulties in diseases of the liver, biliary passages, and pancreas). *Revue Neurologique*, 122(4):263-271, 1970.

In a study of 372 patients suffering from various hepatobiliary diseases, numerous clinical pictures were isolated. Pseudoneurotic states, including pseudoneurasthenia, pseudohysteria, and hypochondriasis, were manifested in a total of 14 Ss, meningeal syndromes in 21, encephalopathy in 26, myelopathy in 13, encephalomyelopathy in 15, visceral autonomic syndromes in 14, and polyneuritis in 27. The most prevalent symptomatology, cerebrovascular syndromes, was present in 67 Ss. The characteristic trait of each of these syndromes was found to constitute a direct evolutionary tie with the typical manifestations of hepatobiliary diseases. Biochemical, EEG, rheoencephalographic, and anatomopathological examination of the brain confirmed the methodological basis of the contemporary conception of the complexity, diversity, and interdependence of bonds between the viscera and the nervous system, as well as their reciprocal influence within the physiological and pathological state. The physiopathological basis is found in difficulties in different types of metabolisms, especially those of the proteins, lipids, and electrolytes. (No refs.) - *B. J. Grylack.*

Clinique de Neurologie et de Neuro-Chirurgie
de l'Universite de l'Amitie des Peuples
Moscow, USSR

1786 PIERSON, M. L'avenir eloigne des enfants hypothyroidiens (The longterm future of hypothyroid children). *Archives Francaises de Pediatrie*, 27(4):337-340, 1970.

Personal experience with approximately 200 hypothyroid children has indicated that the prognosis is always favorable if the disease is treated adequately and systematically. Elimination of the somatic deficit can be realized in 1 to 2 years, depending upon the age of the child at the onset of therapy. Psychomotor coordination is achieved slowly in these children, in whom character and temperament are connected with an early and constant emotional hypersensibility. Intellectual development varies with the age: in young hypothyroid children, psychomotor performance scales are usually a more reliable index than psychometric tests, while academic progress is the best indicator of intellectual level in older Ss. Although hypothyroidism cannot be detected *in utero* at present, preventive hormone therapy administered to the mother has been found effective. (No refs.) - *B. J. Grylack.*

C. H. U.
54 - Nancy, France

1787 ROYER, P.; BALSAN, S.; BROYER, M.; & HABIB, R. Facies d'hypercalcemie idiopathique et hypoplasie segmentaire unilaterale du rein (Physiognomy of idiopathic hypercalcemia and unilateral segmental hypoplasia of the kidney). *Archives Francaises de Pediatrie*, 27(1):19-29, 1970.

A 12-year-old girl presented with idiopathic hypercalcemia associated with segmental hypoplasia of the left kidney with arterial hypertension. Family history revealed that the maternal grandparents of the S and the maternal grandmother of her mother had either suffered from or died of arterial hypertension. No hypercalcemia or hypersensitivity to vitamin D was evident, and she manifested neither osteosclerosis nor nephrocalcinosis. EEG, sex chromatin, and karyotype were normal. The child had an IQ of 90 and manifested the characteristic idiopathic hypercalcemic features of thick lips and large mouth, small nose, and slight hypertelorism. The left superior polar nephrectomy induced a decrease in arterial hypertension, with a rise in blood pressure occurring 18 months following the operation. This association of idiopathic hypercalcemia with segmental hypoplasia of the left kidney with arterial hypertension represents a new cause of renovascular hypertension connected with the characteristic physiognomy and is further support for the primitive anomaly of intrarenal arteries in the origin of segmental hypoplasia. (29 refs.) - *B. J. Grylack.*

Hopital des Enfants-Malades
75 - Paris 15^{e}, France

1788 ROMEO, GIOVANNI; & MIGEON, BARBARA RUBEN. Genetic inactivation of the α-galactosidase locus in carriers of Fabry's disease. *Science*, 170(3954):180-181, 1970.

Fibroblasts from an 18-year-old male with Fabry's disease and his family were studied by colorimetric assay to determine the genetic pattern of the disease. Fabry's disease is an X-linked inborn error of metabolism characterized by the abnormal accumulation in several tissues of a neutral glycolipid, ceramidetrihexoside. Fibroblasts from the affected male showed low α-galactosidase activity in comparison with controls. Study of fibroblasts from the mother and sister showed inactivation of the α-galactosidase locus in female somatic cells and evidence that both were carriers of the disease. The activity of α-galactosidase can also be determined in mixed populations of fibroblasts and epitheloid cells from amniotic fluid. Since α-galactosidase activity is deficient in the fibroblasts of the patient, the assay used in this study can be employed for the prenatal diagnosis of Fabry's disease. (24 refs.) - *B. J. Grylack.*

Johns Hopkins University School of Medicine
Baltimore, Maryland

1789 STRAUSS, RONALD G.; MOHAMMED, SHAKIL; LOGGIE, JENNIFER M. H.; SCHUBERT, WILLIAM K.; FASOLA, ALFRED F.; & GAFFNEY, THOMAS E. The effect of methyldopa on plasma renin activity in a child with Bartter's syndrome. *Journal of Pediatrics*, 77(6):1071-1074, 1970.

A moderately MR boy with Bartter's syndrome showed a decrease in plasma renin activity when treated with methyldopa. The marked increase in circulating renin in response to tilting to a head-up position was also suppressed. The long-term

effects on electrolytes, aldosterone, and psychomotor development require further investigation. (5 refs.) - *E. L. Rowan.*

University of Cincinnati College of Medicine
Cincinnati, Ohio 45219

1790 **OKADA, SHINTARO; VEATH, M. LOIS; & *O'BRIEN, JOHN S.** Juvenile GM_2 gangliosidosis: Partial deficiency of hexosaminidase A. *Journal of Pediatrics,* 77(6):1063-1065, 1970.

Partial deficiencies of hexosaminidase A were noted in 2 patients with juvenile GM_2 gangliosidosis. The syndrome of cerebral accumulation of ganglioside with progressive mental and motor deterioration appears to be a late variant of Tay-Sachs disease where the enzyme is absent and the consequent ganglioside accumulation massive. Both parents of one case had intermediate levels of hexosaminidase A in serum and skin fibroblasts, suggesting an autosomal recessive inheritance of the disorder. (7 refs.) - *E. L. Rowan.*

*University of California, San Diego
La Jolla, California 92037

1791 **LACEY, STEPHEN H.; EYRING, EDWARD J.; & SHAFFER, THOMAS E.** Pycnodysostosis: A case report of a child with associated trisomy X. *Journal of Pediatrics,* 77(6):1033-1038, 1970.

An 18-year-old girl with clinical evidence of pycnodysostosis was incidentally noted to have trisomy X. Short stature, increased bone density, facial and dental anomalies, hypoplastic terminal phalanges, and a tendency to bone fractures were noted. Intelligence is in the low-normal range. Studies showed both decreased calcium accretion and decreased calcium resorption. The syndrome is believed to result from autosomal recessive transmission and is consequently incidental to the trisomy. (9 refs.) - *E. L. Rowan.*

Children's Hospital
Columbus, Ohio 43205

1792 **ETO, YOSHIKATSU; & KITAGAWA, TERUO.** Wolman's disease with hypolipoproteinemia and acanthocytosis: Clinical and biochemical observations. *Journal of Pediatrics,* 77(5):862-867, 1970.

In addition to the vomiting, steatorrhea, dehydration, hepatosplenomegaly, and adrenal calcification characteristic of Wolman's disease, a Japanese infant also showed hypolipoproteinemia and acanthocytosis of red blood cells. There was an extraordinary accumulation of triglycerides, diglycerides, and cholesterol esters in liver and spleen. The stroma of red cells showed low cholesterol and total lipids. Quantitatively there were some abnormalities in these lipids. Brain lipid analysis was suggestive of disordered myelin formation. The total picture is suggestive of a disorder of lipoprotein metabolism in this child. (14 refs.) - *E. L. Rowan.*

University of Pennsylvania Hospital
Philadelphia, Pennsylvania 19104

1793 **SCOTT, C. RONALD; CLARK, SANDRA H.; TENG, CECILIA CHIANG; & SWEDBERG, KATHRYN R.** Clinical and cellular studies of sarcosinemia. *Journal of Pediatrics,* 77(5):805-811, 1970.

A 2-year-old MR boy was found to have excess sarcosine in urine and blood plasma. Sarcosine is an intermediate in "one-carbon metabolism" and is converted to glycine by sarcosine dehydrogenase. Sarcosine loading resulted in very high levels in this patient. Attempts to assay sarcosine or sarcosine dehydrogenase in cultured leukocytes or fibroblasts were unsuccessful even in normal patients, suggesting that this amino acid cannot be utilized *in vitro*. Confirmation of an enzyme deficiency in sarcosinemia requires direct assay of liver tissue from an affected patient. (16 refs.) - *E. L. Rowan.*

University of Washington
Seattle, Washington

1794 **BERMAN, JULIAN L.; & FORD, ROBIN.** Intelligence quotients and intelligence loss in patients with phenylketonuria and some variant states. *Journal of Pediatrics,* 77(5):764-770, 1970.

Children with phenylketonuria (PKU) treated with a low phenylalanine diet were found to

have low-normal intelligence but scored lower on IQ tests than would be expected within their families. Of the 64 children from 48 families, 33 were considered to have PKU (serum phenylalanine greater than 20 mg%), 24 to be PKU variants (levels between 6 and 20 mg%) and 7 to be transient variants (levels less than 6 mg%). Intelligence tests were given to all subjects, siblings, and parents, and a multiple regression equation was developed to estimate the expected level of intelligence within each family. Children with transient or variant PKU scored within the normal range and as expected for their families; however, the children with PKU showed an IQ loss even though scoring within the normal range. Family factors must always be considered in studies such as this. (29 refs.) - *E. L. Rowan.*

Cook County Hospital
Chicago, Illinois 60612

1795 HABERLAND, CATHERINE; & BRUNN-GRABER, ERIC G. Early infantile neurolipidosis with failure of myelination: A chemical and histopathological study. *Archives of Neurology*, 23(6):481-488, 1970.

A case report is presented of a female child who demonstrated early infantile neurolipidosis and a failure of myelination; biochemical and histopathologic data are featured. Details are given in support of the thesis that there is a common biochemical etiology for both the lack of myelin synthesis and abnormal neuronal lipid metabolism. Possibly the earlier synthesis of abnormal glycosides and gangliosides adversely affected myelin synthesis. (31 refs.) - *E. Kravitz.*

Illinois State Psychiatric Institute
Chicago, Illinois

1796 CHOU, S. M.; & THOMPSON, HARTWELL G. Electron microscopy of storage cytosomes in Kufs' disease. *Archives of Neurology*, 23(6):489-501, 1970.

A case of adult cerebral lipidosis (Kufs' disease) is presented. Two characteristic storage cytosomes of the neurons were observed by electron microscopy; lipofuscin-lipid compound bodies and miniature membranous cytoplasmic bodies. There was a generalized increase of ganglioside in the cerebral cortex. Progressive psychomotor retrogression resulted in death. (39 refs.) - *E. Kravitz.*

West Virginia University School of Medicine
Morgantown, West Virginia

1797 MYRIANTHOPOULOS, NTINOS C.; NAYLOR, ALFRED F.; & ARONSON, STANLEY M. Tay-Sachs disease is probably not increasing. *Nature*, 227(5258):609, 1970. (Letter)

The apparent biologic advantage of heterozygotes (4.6-5.3%) over normal homozygotes for the Tay-Sachs gene, as suggested by observations on Eastern European Jews, seems not to be demonstrable in Ashkenazi Jews in North America, Britain, or Israel. The ecologic factors formerly present in Polish ghettos no longer seem to prevail. (3 refs.) - *E. Kravitz.*

National Institute of Neurologic
Disease & Stroke
Bethesda, Maryland

1798 DE GEORGE, FRANCES V. Maternal thyroid disease and twinning. *Nature*, 228(5272):681, 1970. (Letter)

As a result of a study on 409 mothers of twins, it is suggested that thyroid dysfunction may result in increased twinning. The excess of women with twins (over controls with single births) who had thyroid pathologies is statistically significant, e.g., $p<0.05$. Increased thyroid disease and/or thyroid antibodies in mothers have been associated with chromosomal aberrations in offspring, e.g., Down's and Turner's syndromes, and the twinning rate is high among mothers of children with Turner's syndrome. (17 refs.) - *E. Kravitz.*

University of Wisconsin
Madison, Wisconsin

1799 DAVIS, STARKEY D.; NELSON, THOMAS; & SHEPARD, THOMAS H. Teratogenicity of vitamin B_6 deficiency:

Omphalocele, skeletal and neural defects, and splenic hypoplasia. *Science*, 169(3952):1329-1330, 1970.

Pregnant rats were treated with a deficient diet and a B_6 antagonist, 4-deoxypyridoxine, in order to create a state of vitamin B_6 deficiency to be used as an experimental model for a teratologic system exhibiting combined immunologic and neurologic or skeletal defects in humans. As a result of the imposed conditions, the rat fetuses were decreased in size, anemic, and exhibited malformations including omphalocele, exencephaly, cleft palate, micrognathia, digital defects, and splenic hypoplasia. Vitamin B_6 deficiency inhibits both cellular and humoral immunity in experimental animals. Surprisingly, the thymus appeared grossly normal in this study, despite expectation of severe lymphoid hypoplasia. It is anticipated that fetal B_6 deficiency will cause a permanent immunologic defect. (10 refs.) - *B. J. Grylack.*

University of Washington School of Medicine
Seattle, Washington 98105

1800 CHOI, KYOO W.; & BLOOM, ARTHUR D. Biochemically marked lymphocytoid lines: Establishment of Lesch-Nyhan cells. *Science*, 170(3953):89-90, 1970.

A 2-year-old white male with Lesch-Nyhan syndrome marked by MR, compulsive self-mutilation, and hyperuricemia yielded blood from which 2 lymphocytoid cell lines were established and were found to be deficient in hypoxanthine-guanine phosphoribosyltransferase (HGPRT) activity. This was the first time that a presumably permanent human lymphocytoid cell line was shown to be deficient in a specific enzyme. The technique based on the mechanism by which lysates of previously established lymphocytoid cell lines enhance establishment of new, presumably permanent lines has a higher success rate than repeated stimulation with phytohemagglutinin and a decreased requirement for amount of blood needed from the patient for initiation of cell cultures. The establishment and maintenance of permanent lymphocytoid cell lines may provide an excellent alternative system to the fibroblast for the study of genetically determined biochemical defects in human cells. (11 refs.) - *B. J. Grylack.*

University of Michigan Medical School
Ann Arbor, Michigan 48104

1801 Inborn terror of metabolism. *Journal of the American Medical Association*, 214(12):2186-2187, 1970. (Editorial)

The Lesch-Nyhan syndrome results from an X-linked inborn error of metabolism and is characterized by uric aciduria, MR, choreoathetosis, spasticity, and self-mutilation. Perpetuated as a sex-linked recessive trait and mediated by a deficiency of hypoxanthine-guanine phosphoribosyltransferase (HGPRT), which results in purine synthesis, the disease leads to early death before the age of puberty. Due to the failure of known treatments to cure or improve the severe neurologic symptoms, the alternative is early termination of pregnancy if a prenatal diagnosis of an affected male fetus can be established. Amniotic fluid from transabdominal amniocentesis at approximately the eighteenth week of gestation may be used for examination and culture. Screening is best achieved by identifying MR children with significant uric aciduria and by then examining the mother and other female relatives at risk. (2 refs.) - *B. J. Grylack.*

1802 KELLY, SALLY; KATZ, SELIG; BURNS, JEAN; & BOYLAN, JEANNE. Screening for galactosemia in New York State. *Public Health Reports*, 85(7):575-578, 1970.

Screening for phenylketonuria and galactosemia of 141,402 infants in New York State during 1968 and 1969 revealed a galactosemia incidence of approximately 1 in 35,000. A fluorescent spot method of testing proved a practical means of identifying those infants in whom galactosemia was likely to develop. The method indicated activity of galactose-1-phosphate uridyl transferase by development of a fluorescent spot on filter paper after incubation of dried blood sample with substrate. 132 of 143 infants whose samples did not fluoresce were tested a second time, and 110 were considered normal. Of the 22 remaining, third samples were not obtained from 6, 9 were classifed as normal on the third sample, and 7 were classified as afflicted or as carriers. Four of the 7 who exhibited persistent transferase deficiencies were homozygotes. (21 refs.) - *J. K. Wyatt.*

New York State Department of Health
Albany, New York

1803 YU, J. S.; ADAMS, B.; & O'HALLORAN, MARY T. Studies in the tyrosine metabolism of phenylketonurics. *Australian Paediatric Journal*, 6(4):215-217, 1970.

Abnormality in the metabolism of exogenous tyrosine in phenylketonuria was not demonstrated in oral l-tyrosine tests performed on 6 normal control Ss, 11 parents of known classic phenylketonuric children, and 10 classic phenylketonuric children. All phenylketonuric children were on some form of diet. Four who were diagnosed late, MR, and on a modified diet had fasting serum levels >10 mg/11 ml. The other 6 had serum levels <10 mg/11 ml. 100 mg/kg body weight of l-tyrosine was administered orally after an overnight fast. Venous or capillary blood was collected at 0, 1, 2, and 4 hours. Phenylketonuric children exhibited lower values of serum tyrosine than controls; however, this finding reflected lower baseline values. (13 refs.) - *J. K. Wyatt.*

Royal Alexandria Hospital for Children
Sydney, NSW, Australia 2050

1804 HASHEM, NEMAT; EBRAHIM, ASSMA; & NOUR, ASSMA. Classical and typical phenylketonuria among Egyptians: Study of 10 families. *American Journal of Mental Deficiency*, 75(3):329-335, 1970.

Several Egyptian families, in which at least one member had phenylketonuria (PKU), exhibited discrepancies in the degrees of hyperphenylalaninemia, ketonuria, and mental deficiency. Ten Egyptian families including 24 phenylketonuria patients were studied. The families tended to aggregate in the Lower Nile Valley, and 9 families presented overt examples of classical PKU. Dermatoglyphic findings considered common for PKU patients were not observed in the presented cases. The results indicated that the gene prevalence of classical PKU among Egyptians is 0.0067, with a carrier rate for the general population of 1 in 76. PKU Ss with normal intelligence were found, and one case of hyperphenylalaninemia with reduced intelligence presenting as atypical PKU was found. The fact that parents and grandparents of most of the propositi derive from the Suez Canal region indicates admixture of the PKU gene from intermarriage with European invaders. (12 refs.) - *S. Glinsky.*

5, El Goumhouria Street
Cairo, Egypt, U.A.R.

1805 BIRCH, HERBERT G. Functional effects of fetal malnutrition. *Hospital Practice*, 6(3):134-136, 141-144, 147-148, 1971.

Nutritional deprivation early in life and continued into maturity and pregnancy has cumulative consequences for offspring, presaging deficient prenatal growth likely to persist after birth. Associated with fetal malnutrition are deficits in number and size of brain cells and extensive myelination, which are reflected in later mental and behavioral functioning. A variety of studies show low-birthweight infants may suffer gross neurologic damage, measurable intellectual handicaps, and aberrations in perception, abstract reasoning, and attention span. (Testing with Bender-Gestalt and Wechsler has substantiated this.) In addition, prematures are more subject to personality disturbances. Studies in Aberdeen, Scotland, have shown obstetric and perinatal antecedents, including low birthweight, associated with mental subnormality. Early nutrition affects neurointegrative organization, upon which the emergence of complex capacities and sensory interrelationships depends. Studies in rural Mexico have shown sharp differences between urban and rural children, and confirmed the effects of early malnutrition, and of familial and social factors, on mental and adaptive development. There is evidence that malnutrition among U.S. poor is affecting child growth and mental development. Parent education and widespread public health programs are imperative. (No refs.) - *B. Berman.*

Albert Einstein College of Medicine
New York, New York

1806 Inborn errors of metabolism. *Clinical Pediatrics*, 10(1):1-3, 1971.

Recent years have seen rapid increases in the number of inborn errors of metabolism and of affected individuals, with the greatest attention going to PKU and hyperphenylalaninemia, which derive from the multiple allelic mutant forms of the genes controlling the structure of phenylalanine hydroxylase. That multiple forms of this unstable enzyme will emerge rests theoretically

on current knowledge of the glycolytic enzyme, glucose-6-phosphate dehydrogenase, which exists in at least 49 mutant forms, chiefly affecting severe loss of activity in chronic nonspherocytic hemolytic anemia, and with differences occurring among Negroes, Sephardic Jews, Greeks, and Sardinians. These diverse forms serve as a guide for studies of metabolic errors and other recessively inherited diseases. The group of mutant enzymes whose activity can be restored by cofactor is of practical importance, as are the cases of methylmalonic acidemia responding to vitamin B_{12}. Pediatricians must understand the various reasons for the variability seen in PKU and hyperphenylalaninemia, and must be guided by the unfortunate experiences with these metabolic errors, where, despite better identification with the Guthrie test, overenthusiastic dietary treatment has caused many tragic deaths. (13 refs.) - *B. Berman.*

1807 BRENCHLEY, MAUREEN. The child with Hurler's syndrome. *Canadian Nurse*, 67(2):38-39, 1971.

The child with Hurler's syndrome needs tender, loving care, palatable and nourishing food given slowly in small amounts, and sensory stimulation as the concomitant deafness progresses and the vision dims. Relatively rare, the syndrome—an inherited mucopolysaccharide-storage disease involving connective tissue and transmitted as an autosomal recessive trait—causes progressive physical and mental deterioration, beginning in infancy and ending in death at age 7-10 years. The disease, manifesting skeletal anomalies, nasal and respiratory congestion, neck and chest deformities, and contractures, is diagnosed by family history, abnormal X-ray findings, and presence of specific mucopolysaccharides. There is no known cure. Parents require counseling and moral support. The child should be encouraged to be independent: within his physical and mental limitations, he can be taught to use the toilet, dress and feed himself, and wash and clean his body. (3 refs.) - *B. Berman.*

No address

1808 RAINE, D. N. Defects in renal tubular reabsorption of amino acids. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 43-52.

Transport processes across membranes into and out of cells exist and can malfunction or fail completely with frequently serious disease consequences. Of 5 group transport systems for amino acids (basic and cystine, acidic, neutral, imino acids and glycine, beta), abnormalities in humans have been shown in all but the third. Interferences occur with tubular transport, leading to various degrees of amino aciduria. Understanding the transport system helps explain the amino aciduria in specific inherited metabolic disorders that cause accumulation in the blood of a single amino acid. Any condition that damages the renal tubular epithelium affects its transport functions and causes amino aciduria. Diseases associated with such a renal aciduria include a vitamin-D deficiency leading to rickets, Fanconi syndrome, Wilson's disease, and galactosemia. Specific disorders of tubular reabsorption include: Hartnup disease, cystinuria, and iminoglycinuria. There are inherited metabolic diseases (hyperprolinemia, β-alaninemia, citrullinuria, and argininemia) in which the renal tubule becomes overloaded when a single amino acid accumulates in the blood and is filtered by the glomerulus. Therapy for various metabolic disorders has involved, through dietary and other means, the saturation or blocking of transport systems. (36 refs.) - *B. Berman.*

Children's Hospital
Birmingham, England

1809 JEPSON, JOHN B. Hartnup disease. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 55-63.

In Hartnup disease, a genetic, autosomal recessive disorder, a red scaly rash on exposed parts, cerebellar ataxia, and occasional episodes of mental instability, occur. Urinary amino-acid chromatography reveals an extraordinary amino-acid excretion pattern, showing faulty renal reabsorption of the monoamino monocarboxylic acids; renal clearances for the amino acids involved are very high. Of the over 40 cases reported, half have had reversible neurological or neuropsychiatric symptoms. Only 9 are described as MR; 1 is

mentally defective. The renal defect causes some amino-acid losses, but they are not nutritionally significant. The time course for urinary excretion of indican and indolyacetic acid after oral tryptophan indicates a gut defect causing intestinal retention of the free amino acid and unavailability of much of the dietary intake of the essential tryptophan. Several researchers have suggested an endogenous "toxic" agent as cause of the neuropsychiatric signs. (11 refs.) - *B. Berman.*

Middlesex Hospital Medical School
London, England

1810 DINGLE, J. T. Lysosomes in nervous tissue. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 67-73.

Lysosomes are organelles important in the physiology and pathology of almost all cells and tissues. The same immunoenzymic techniques that are explicating lysosomal actions in connective-tissue metabolism are applicable to studying lysosomal function in axon endings. Light and electron microscopy have identified the presence of lysosomes in normal tissue and shown their close relationship to the Golgi apparatus in neurons. Biochemical studies have shown that lysosomal properties are not very dissimilar from those of other tissues. These studies have sought also to explain the lysosomal role in neural pathology (increased levels of lysosomal enzymes are found in cerebrospinal fluid in acute demyelinating disease, diabetic neuropathy, Barre syndrome, and meningeal carcinomatosis), and in the storage diseases (lipidoses and polysaccharidoses) such as Tay-Sachs, Niemann-Pick, Jakob-Creutzfeld, and Alzheimer's diseases. Studies of the lysosomal enzyme in the latter pathologies by immunoenzymic methods would probably yield valuable data. (20 refs.) - *B. Berman.*

Strangeways Research Laboratory
Cambridge, England

1811 MUIR, HELEN. The biochemistry of the mucopolysaccharidoses. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 75-93.

Despite a great deal of research, the biochemical defect in the mucopolysaccharidoses remains unexplained. Characterized by skeletal defects and progressive MR, this group of recessive storage diseases shows a large output of urinary sulphated mucopolysaccharides or GAG (glycosaminoglycans), with indications that the latter derive from the liver and spleen. This group shows striking resemblances to other storage diseases with a lysosomal hydrolase deficiency, and a search for such a deficiency has been strengthened by reports of partial β-galactosidase deficiencies in Hurler, Hunter, and Sanfilippo syndromes (there are various genetic and other theoretical as well as technical difficulties in accepting this explanation). Whatever the nature of the crucial factor, in most mucopolysaccharidoses it affects the metabolism of either dermatan sulphate or heparan sulphate. And, whatever the genetic defects, they primarily affect the biosynthesis of GAG, for which prior formation of the core protein is necessary. Fibroblast cultures have been used with little success to explain the abnormal urinary GAG. The connective-tissue abnormalities may have causes similar to those producing gangliosidosis in pseudo-Hurler's disease. The progressive MR in these diseases is probably caused by the accumulation in the brain of gangliosides and GAGs. (116 refs.) - *B. Berman.*

Kennedy Institute of Rheumatology
Hammersmith, London, England

1812 POLLITT, R. J.; & JENNER, F. A. Aspartylglycosaminuria—A new inborn error possibly due to a lysosomal enzyme defect. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 95-101.

Aspartylglycosaminuria is an inborn metabolic error characterized by manic phases and a biochemical deficiency of AADG (2-acetamido-1-β′-L-aspartamido-1, 2-dideoxy-β-D-glucose) that causes urinary excretion of glycoasparagines. Displaying some storage characteristics, this pathology falls between the aminoacidurias and the lysosomal storage diseases. The storage aspects involve traces of various high molecular-weight compounds in the cerebrospinal fluid in a pattern different from the urinary one. The excessive AADG could be due to a defect in glycoprotein

synthesis or in degradation. There is evidence that a major lysosomal enzyme is missing in this disease. (16 refs.) - *B. Berman.*

Middlewood Hospital
Sheffield, England

1813 **ALLISON, A. C.** Types of lysosomal abnormality and the light they throw on normal lysosomal functions. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 107-114.

The structure and function of lysosomes are affected by 3 categories of congenital abnormality: (1) lysosomal-enzyme defect (failure of normal lysosomal-enzyme synthesis); (2) Chediak-Higashi syndrome (found in individuals homozygous for an autosomal mutant gene, it shows unusually large granules in peripheral-blood leukocytes and dilution of skin, hair, and eye color, and produces a susceptibility to various infections and malignancies); and (3) chronic granulomatous disease of children (defective antimicrobial action of certain lysosomes). Inclusion bodies, in the first abnormality, accumulate because of a missing enzyme. This reflects a class of abnormalities in which unstable proteins are secondarily incorporated into lysosomes. The role of lysosomes in protein synthesis and turnover is important, and may be associated with abnormal neuronal functioning. (35 refs.) - *B. Berman.*

Northwick Park Hospital
Harrow, England

1814 **MYANT, M. B.** The biochemistry of the gangliosides in relation to inborn disorders of ganglioside metabolism. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 117-131.

Gangliosides (G) are categorized under the complex lipids known as sphingolipids. Although very little is known about their physiologic role, their high concentration in brain (at least 5 enzymes concerned in ganglioside breakdown have been isolated from brain tissue) suggests that they affect specialized nerve-tissue function. Gangliosides seem to belong to a larger group of sphingolipidoses (these include Niemann-Pick, Fabry's, and Gaucher's diseases), in which there is an inherited deficiency of lysosomal acid hydrolase. Inborn metabolic errors may cause defects in biosynthetic pathways and excessive ganglioside accumulations such as occur in Tay-Sachs disease. In the more usual form of Tay-Sachs disease, the brain accumulation of gangliosides seems due to a deficiency of β-hexosaminidase, although several important questions remain unanswered. Several metabolic disorders are due to a β-galactosidase deficiency. These include the familial disease known as generalized gangliosidosis and Fabry's disease (glycolipid lipidosis). The rare infantile form of Gaucher's disease is not a true gangliosidosis, but shows brain cerebroside as largely replaced by glucocerebroside. Generally, in all these disorders, a defect in a catabolic pathway causes a pathologic accumulation of a normal intermediate; where an enzyme causes the block, it appears to be an acid hydrolase associated with the lysosomes. (31 refs.) - *B. Berman.*

Hammersmith Hospital
London, England

1815 **RAINE, D. N.** Laboratory diagnosis of some sphingolipidoses. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 133-139.

Chemical techniques, such as thin-layer chromatography and sophisticated enzyme assays, are replacing brain biopsy or necropsy in diagnosing various sphingolipidoses (gangliosidosis, Gaucher's disease, globoside storage disease, Fabry's disease, metachromatic leukodystrophy). Since these diseases involve inherited enzyme deficiencies relevant to complex lipid metabolism, they may be studied by assaying the enzyme or seeking signs of abnormal lipid accumulations. Diagnostic methods applicable to one or more of these diseases include checking for: circulating leukocyte enzyme deficiency, urine enzyme activity, plasma enzyme activity, enzyme activity in cultured fibroblasts, abnormal lipids in urine sediment, and enzyme activity in amniotic-fluid cells. (25 refs.) - *B. Berman.*

Children's Hospital
Birmingham, England

1816 **SEEGMILLER, J. EDWIN.** Lesch-Nyhan syndrome: Preventive control. *Science*, 170(3964):1333, 1970.

In preventive control by prenatal diagnosis of Lesch-Nyhan syndrome, the diagnosis of index cases has practical significance, since it leads to the identification of heterozygotic mothers who are at risk of bearing additional affected children and to the examination of other female relatives for evidence of cell mosaicism for the enzyme in question. Despite the theoretical consideration that new mutations not reflected in the somatic cells of the mother could account for as many as a third of all cases, mosaicism was found in all 10 mothers examined by the laboratory. (1 ref.) - *B. J. Grylack.*

University of California
San Diego, California

1817 **THIER, SAMUEL O.** Guilt by association? *New England Journal of Medicine*, 283(15):815-816, 1970. (Editorial)

Although cystinuria and mental disease have been linked in a recent study, this association cannot be accepted as firmly established. Cystinuria manifested clinically by calculi in the urinary tract results from a defect in transepithelial transport of cystine and the dibasic amino acids—lysine, arginine, and ornithine. This defect, expressed in the epithelial cells of the renal tubule and the gastrointestinal tract, is inherited as a recessive trait with 2 forms of expression in the heterozygous state: the completely and incompletely recessive forms of cystinuria. Defective intestinal absorption and increased renal clearance of cystine and the dibasic amino acids could produce mental dysfunction through several mechanisms. In view of the fact that cystine has been established as an essential amino acid for the brain in early life and that cow's milk is relatively high in methionine and low in cystine, a cystinuric child unable to absorb cystine and fed cow's milk might have impaired cerebral metabolism. The connection between the transport defect of cystinuria and the presence of mental disease can be constructed, but the data underlying this link must be evaluated with an understanding of the difficulty entailed in determining the prevalence of homozygous cystinuria in the general population and with a sufficiently large abnormal population sample. (5 refs.) - *B. J. Grylack.*

No address

1818 **TAPIA, HUGO R.; & CAVAZOS, NINFA.** Hepatolenticular degeneration (Wilson's disease): Observations in 7 cases and review of the literature. *Southern Medical Journal*, 63(1):82-89, 1970.

Seven cases of hepatolenticular degeneration (Wilson's disease) were observed over a 12-year period at a hospital center. This disease appears to be a result of abnormal copper metabolism and is manifested by dysarthria, dysphagia, muscle hypertonicity, tremor, and dyspraxia together with the characteristic Kayser-Fleischer ring. Wilson's disease is diagnosed primarily in young persons and is based upon an autosomal recessive inheritance pattern. Laboratory tests reveal increased copper in body organs, increased urinary copper, decreased serum copper, and a deficiency of serum ceruloplasmin. Penicillamine is the chelating agent used to form complexes with the copper. Muscle relaxants and anti-parkinsonian drugs have been employed to counteract involuntary movements. Since chelation therapy is significant if extensive damage has not been done, early diagnosis is the key to effective treatment. (20 refs.) - *B. J. Grylack.*

University of Texas Medical Center
Galveston, Texas

1819 **LINES, DAVID R.; & WAISMAN, HARRY A.** Urinary amino acid excretion in phenylketonuric, hyperphenylalaninemic, and normal patients. *Journal of Pediatrics*, 78(3):474-480, 1971.

Statistically significant abnormal patterns of renal excretion of amino acids were found in 11 phenylketonurics who were off low-phenylalanine diets, 7 phenylketonurics who were on low-phenylalanine diets, and 7 hyperphenylalaninemic children when compared with 7 normal children. The pattern of amino acid excretion in off-diet phenylketonurics was suggestive of competitive inhibition of reabsorption of amino acids by the high filtered load of phenylalanine. A generalized aminoaciduria found in hyperphenylalaninemic

patients suggested that they may be phenylketonuric individuals who are protected by increased urinary excretion of phenylalanine. On-diet phenylketonuric patients exhibited generalized aminoaciduria which was probably related to their highly modified diet. (21 refs.) - *J. K. Wyatt.*

University of Wisconsin Medical Center
Madison, Wisconsin 53706

1820 **CHACKO, CLARAMMA M.; CHRISTIAN, JOE C.; & NADLER, HENRY L.** Unstable galactose-1-phosphate uridyl transferase: A new variant of galactosemia. *Journal of Pediatrics,* 78(3):454-460, 1971.

A new variant of clinical galactosemia, the Indiana variant, appears to be characterized by the presence of an additional allele at the transferase locus. A female patient, CA 18 mos, presented evidence of storage instability of transferase in heparin and in phosphate buffer and an altered electrophoretic mobility. Galactose-1-phosphate uridyl transferase activity in the red blood cells was approximately 40% of normal. Treatment was by a galactose-restricted diet, and growth and development have since been within normal limits. Both the mother and maternal grandmother demonstrated storage instability of galactose-1-phosphate uridyl transferase and electrophoretic mobility that was slower than normal. Genetic studies indicated that the patient appeared to be a double heterozygote with the 'classic' galactosemia gene inherited from the father and the Indiana variant galactosemia gene inherited from the mother. The variant gene may have resulted in the synthesis of a structurally altered protein in which changes in the 3 dimensional configuration of the enzyme caused molecular instability and interference with its function. (16 refs.) - *J. K. Wyatt.*

Children's Hospital
Chicago, Illinois 60614

1821 **KATTAMIS, CHRISTOS; KARAMBULA, KATERINA; METAXOTOU-MAVROMATI, ANNA; & MATSANIOTIS, NICOLAS.** G-6-PD deficiency and age. *Lancet,* 1(7692):235, 1971. (Letter)

Screening for G-6-PD deficiency in Rhodes, Greece, of 619 boys and adolescents (CA 6-20 yrs) and 100 MR males revealed that 95% of MRs had values ranging from 0-5% of the normal mean. Five percent of MRs had intermediate level values (17-60%) which corresponded to those found in the mild Mediterranean variant. No significant differences in the frequencies of G-6-PD deficiencies or β-thalassemia trait were found among normal Ss in the age groups 6 to 10 years, 11 to 15 years, and 16 to 20 years. (2 refs.) - *J. K. Wyatt.*

University of Athens
Athens, Greece (608)

1822 **GOODMAN, STEPHEN I.; MACE, JOHN W.; & POLLAK, SHLOMO.** Serum gamma-glutamyl transpeptidase deficiency. *Lancet,* 1(7692):234-235, 1971. (Letter)

A moderately MR male exhibited deficient activity of serum γ-glutamyl transpeptidase. Estimations of total serum and urine glutathione concentrations showed a thousand-fold increase in concentration in the urine as well as a moderate hyperglutathionemia. Measurement of serum γ-glutamyl transpeptidase activity in the patient and 14 control Ss revealed a profound decrease in the activity of the enzyme in the patient. The results of mixing experiments indicated that this decrease was not due to the presence of glutathione. The glutathionemia and glutathionuria observed in this patient may be due to a generalized deficiency of γ-glutamyl transpeptidase activity of unknown etiology. (9 refs.) - *J. K. Wyatt.*

University of Colorado Medical Center
Denver, Colorado 80220

1823 **WESTALL, R. G.; CAHILL, ROSE; & SYLVESTER, P. E.** Hyperglucaricaciduria. A new error in carbohydrate metabolism found in a mentally retarded patient. *Journal of Mental Deficiency Research,* 14(4):347-351, 1970.

An institutionalized male MR (CA 21 yrs; IQ 50) excreted over 5 g of D-glucaric acid/day in his urine. The patient was on a normal hospital diet.

He appeared to have a metabolic block on the glucose-glucuronic acid-pentose pathway. Adjustment to grossly increased amounts of glucaric acid in his system was good. However, since this condition appears to have been lifelong, the high levels of D-glucaric acid in his system may have contributed to MR and retarded physical growth. (7 refs.) - *J. K. Wyatt.*

St Lawrence's Hospital
Caterham, Surrey, England

1824 BLEHOVA, BOHUNKA; PAZOUTOVA, NOEMI; & SUBRT, I. Phenylketonuria associated with Down's syndrome. *Journal of Mental Deficiency Research*, 14(4):274-275, 1970.

Both phenylketonuria and Down's syndrome were presented by an SMR boy (date of birth, 1946; IQ in 1969, 20). Phenylketonuria was confirmed at age 12 when the boy was examined for the first time. Chromosomal analysis revealed 47 chromosomes with 21-trisomy and dermatoglyphs typical of Down's syndrome. Development had been slow, and speech did not develop. The boy did not attend to his environment and sat making salaam movements. This is the second known report of phenylketonuria in association with Down's syndrome. When phenylketonuric children who have been treated with a well-monitored diet from infancy do not develop as expected, MR may be due to a different etiology. (13 refs.) - *J. K. Wyatt.*

Prague 10, Vinohrodska 159,
Czechoslovakia

1825 BENSON, PHILIP F.; BOWSER-RILEY, FRANK; & GIANNELLI, FRANCESCO. β-galactosidases in fibroblasts: Hurler and Sanfilippo syndromes. *New England Journal of Medicine*, 283(18):999-1000, 1970. (Letter)

The activities of β-galactosidases, Types B and C, were at least as high in fibroblasts from subjects with mucopolysaccharidoses (Types 1 to 3) as in normal controls. Normal values in 4 human controls were 1014-1440 units. A 5-year-old girl with Sanfilippo's syndrome had values of 1466-1946 units; a boy with Hurler's syndrome had values of 1016-1916 units. (5 refs.) - *E. Kravitz.*

Guy's Hospital Medical School
London, England

1826 LEROY, JULES G.; & SPRANGER, JURGEN W. I-cell disease. *New England Journal of Medicine*, 283(11):598-599, 1970. (Letter)

Various aspects of I-cell (inclusion-cell) disease (mucolipidosis II) are discussed. The I-cell phenotype is also present in mucolipidosis I. Six additional cases of I-cell disease have been studied since the original article was published. This disease is characterized by severe psychomotor retardation, respiratory infections, short stature, and progressive Hurler-like alterations of facial and body configuration; the condition is an autosomal genetic recessive. (7 refs.) - *E. Kravitz.*

State University
Antwerp, Belgium

1827 CARTON, D.; DACREMONT, G.; KLINT, J.-A.; & VLIETINCK, R. Gangliosidose a G_{M1} type II (G_{M1} gangliosidosis, type II). *Archives Francaises de Pediatrie*, 27(10):1089-1090, 1970.

Profound psychomotor retardation was observed in a 17-month-old male. No familial pathology was revealed. Clinical abnormalities included disappearance of spontaneous motor movements, hypertonia and hyperreflexia, and moderate hyperacousia. Enzymatic study disclosed a pronounced decrease of β-galactosidase activity in the leukocytes and the liver, as well as a moderate decrease in the grey matter of the central nervous system. The presence of the G_{M1} fraction and its axial derivative in the brain but not in the liver, in addition to the clinical symptoms of late onset, long evolution, and the presence of facial dysmorphia, visceromegaly, and ophthalmological and skeletal anomalies, confirmed the diagnosis of G_{M1} gangliosidosis, type II in contrast to generalized G_{M1} gangliosidosis. (No refs.) - *B. J. Grylack.*

No address

1828 **DE SCHRIJVER, F.** Syndrome de Sanfilippo (Sanfilippo's syndrome). *Archives Francaises de Pediatrie,* 27(10):1092, 1970.

A male child aged 4½ years, investigated because of mental development anomalies and characterological difficulties, manifested facial dysmorphia and hepatomegaly. Radiological examination and study of the urinary mucopolysaccharides favored the diagnosis of mucopolysaccharidosis, but clinical and radiological comparison of the data with those present in other metabolic disorders of mucopolysaccharides indicated the probable existence of polydystrophic oligophrenia, or the Sanfilippo syndrome. (No refs.) - *B. J. Grylack.*

No address

1829 **EGGERMONT, E.; BRACKE, P.; EMMERY, L.; VAN DER SCHUEREN-LODEWEYCKX, M.; PROESMANS, W.; TIMMERMANS, J.; VAN HOOF, P.; & EECKELS, R.** Un cas de gargoylisme connatal (A case of congenital gargoylism). *Archives Francaises de Pediatrie,* 27(10):1095, 1970.

A 1-year-old prematurely born male presented with a significant height and weight deficit (58 cm and 4.4 kg), marked psychomotor retardation, bowing of the lower extremities, facial dysmorphia, and pronounced hepatosplenomegaly. Radiological examination revealed severe osteoporosis with irregularities of the bone cortices and medullae. The base of the cranium was very thick. Ophthalmological examination disclosed corneal opacities which had appeared at 5 months of age. A particular type of congenital gargoylism was diagnosed on the basis of immense vacuoles in the lymphocytes of circulating blood and pathological inclusions in the hepatocytes, normal urinary excretion of mucopolysaccharides, inclusion bodies among fibroblasts in culture, and the absence of any particular enzymatic lesion. (No refs.) - *B. J. Grylack.*

No address

1830 **VERGER, P.; CHANNAROND, J.; GUILLARD, J.-M.; & GUITER, F.** Les formes a survie prolongee de la maladie de Hurler. Le probleme du Spat-Hurler (Forms of prolonged survival in Hurler's disease. The problem of the term Spat-Hurler). *Archives Francaises de Pediatrie,* 27(8):864, 1970.

Three cases of late-onset Hurler's disease with prolonged duration have been reported. A 6-year-old boy presented with clear manifestations of the disease in the form of characteristic facial morphology, deafness, and lumbar kyphosis, although MR, corneal opacities, and the typical vertebral radiological anomaly were absent. At age 22 the S revealed marked dwarfism and dysmorphia typical of Hurler's disease, but his intellectual capacity was intact. Two sisters aged 7 and 5 years were diagnosed as suffering from Hurler's disease on the basis of their facial morphology, corneal opacities, and articulatory difficulties. At age 21 and 19 they were dwarfish, kyphotic, deaf, and had very defective vision and claw-like hands. Except for the absence of typical vertebral radiological lesions in the 3 Ss, it would seem impossible to anticipate their prolonged survival. It appears necessary to reserve the term Spat-Hurler's disease for Ss who, as the 2 sisters did, present with a reduced height, prognathism, corneal opacities, and claw-like hands. However, no biochemical criterion differentiates these Ss at present from the autosomal recessive form of classical Hurler's disease. (No refs.) - *B. J. Grylack.*

No address

1831 **TONDEUR, M; VAMOS-HURWITZ, E.; CREMER, N.; & LOEB, H.** Mucopolysaccharidosis in a three months old infant. *Acta Paediatrica Scandinavica,* 60:98-101, 1971.

A case of mucopolysaccharidosis, type I, in a 3 month old female is presented. Diagnosis was based upon the presence of facial dysmorphism, MR, hepatosplenomegaly, inguinal hernia, joint stiffness and corneal clouding, increased mucopolysaccharidiuria affecting heparan and dermatan sulfate, and vacuolized lymphocytes. Unusual features include protruding sternum, high dorsal kyphosis, a normal β-galactosidase activity, anomalous ultrastructure of liver cells, and a rapidly fatal evolution of the disease. (12 refs.) - *N. Jarvis.*

Department of Paediatrics
University of Brussels, Belgium

1832 **EID, E. E.** A follow-up study of physical growth following failure to thrive with special reference to a critical period in the first year of life. *Acta Paediatrica Scandinavica*, 60:39-48, 1971.

Seven parameters of growth — standing height, supine length, weight, head circumference, chest circumference, triceps and subscapular subcutaneous folds, and skeletal age — were tested in a group of children considered to have failure to thrive and in a control group. A treated group consisted of surgical and medical cases in whom the cause of failure to thrive was removed, and the untreated control group consisted of cases in which treatment of the cause was impossible. In six parameters significant retardation was noted in both groups. Within the treated group neither early onset of failure to thrive nor its severity caused severe retardation, but prolonged duration of failure to thrive or failure to correct the cause in the first year led to significant growth retardation. But since it is difficult to separate the effect of illness from that of genetic endowment, this must be taken into consideration in estimating the importance of events in the first year of growth. The smaller size of children with congenital anomalies may in itself be a congenital anomaly independent of the original anomaly. (29 refs.) - *N. Jarvis.*

Sheffield University
Sheffield, England

1833 **HJALMARSSON, O.; JAGENBURG, R.; & RODJER, S.** Mild and severe PKU. Comparative studies in two infants. *Acta Paediatrica Scandinavica*, 60:11-16, 1971.

Changes in plasma phenylalanine level during fasting periods, after meals, and after intravenous phenylalanine infusion were examined for differences in phenylalanine turnover in two infants, one with classical severe and one with mild phenylketonuria (PKU). Case reports, methods, and results are described, and classifications of types of PKU are discussed. The results obtained revealed no significant differences in turnover rate. (24 refs.) - *N. Jarvis.*

University of Gothenburg
Gothenburg, Sweden

1834 **SKREDE, SVERRE; STROMME, JOHAN H.; STOKKE, ODDVAR; LIE, SVERRE O.; & ELDJARN, LORENTZ.** Fatal congenital lactic acidosis in two siblings. II. Biochemical studies *in vivo* and *in vitro*. *Acta Paediatrica Scandinavica*, 60:138-145, 1971.

In vivo and *in vitro* studies of one of two infants who later died of congenital lactic acidosis were performed in an attempt to localize the metabolic defect. Estimations of radioactive and cold lactate, pyruvate, and glucose in serum; total CO_2 and $^{14}CO_2$; urinary lactate; glycogen content of tissues; lactic dehydrogenase (LDH) isoenzymes; and liver and muscle metabolism were made. Possible causes for the accumulation of lactate include an enzyme defect, a toxic effect of an accumulated metabolite, an increase in lactate production, a decreased oxidation of lactate, and a block in the gluconeogenesis. Abnormally high production of lactate was excluded as a hypothetical cause by the results of the *in vivo* study; this exclusion indicates that a reduced capacity for lactate removal exists. Comparative estimates revealed an abnormally low capacity for lactate removal in the patient. No insufficient capacity for the oxidation of lactate/pyruvate was revealed, and oxidation accounted for about 60% of estimated lactate removal. On the other hand, there was substantial evidence for a block in the gluconeogenetic pathway: a very low glycogen content in both liver and muscle; negligible amounts of radioactive glycogen in the liver and of radioactivity in blood glucose; evidence that recycling of radioactivity from glucose did not occur. Certain discrepancies between this study and others, pointing to a functioning gluconeogenesis as a cause of hyperlactacidemia, suggest that congenital lactic acidosis might be a term for a heterogeneous group of diseases. (33 refs.) - *N. Jarvis.*

University of Oslo
Rikshospitalet, Oslo, Norway

1835 **PALLISGAARD, G.; & GOLDSCHMIDT, E.** The oculo-cerebro-renal syndrome of Lowe in four generations of one family. *Acta Paediatrica Scandinavica*, 60:146-148, 1971.

The oculo-cerebro-renal syndrome described by Lowe is investigated in 4 generations of a Danish

family, with particular reference to genetic analysis. Transmission of the syndrome was sex linked, all affected boys showing typical features, such as cataract/glaucoma, psychomotor retardation, muscular hypotonia, and renal manifestations. All carriers had lenticular opacities, ranging from coerulea elements to peripheral spokes to posterior polar cataract. Ophthalmologic examinations are recommended for all possible carriers of the Lowe syndrome prior to genetic counseling. (10 refs.) - *N. Jarvis.*

Odense University Hospital
Odense, Denmark

1836 **SUZUKI, YOSHIYUKI; CROCKER, ALLEN C.; & SUZUKI, KUNIHIKO.** G_{M1}-Gangliosidosis. Correlation of clinical and biochemical data. *Archives of Neurology*, 24(1):58-64, 1971.

Postmortem specimens from 8 patients with G_{M1} gangliosidosis and from normal control patients were assayed to correlate biochemical findings with clinical manifestations. Three criteria were chosen for comparison: G_{M1} ganglioside storage in brain and visceral organs; visceral accumulation of keraton sulfate-like mucopolysaccharide; and β-galactosidase deficiency. Two clinical types of the disease had previously been distinguished, one exhibiting features associated with Hurler's syndrome and a second in which no such features appeared. No one of the 3 biochemical criteria proved sufficient in itself to justify a diagnosis of G_{M1} gangliosidosis: the diagnosis does not necessarily depend upon the presence of brain ganglioside pathology; visceral G_{M1} ganglioside may be normal in type 2 patients; abnormal increase of visceral keraton sulfate-like mucopolysaccharide may occur in other neurological disorders, and analysis of the compound is subject to a wide range of errors; and galactosidase deficiency does not necessarily exclude Hurler-Hunter syndrome. Therefore, all 3 criteria should be used for the diagnosis of G_{M1} gangliosidosis. (40 refs.) - *N. Jarvis.*

University of Pennsylvania School of Medicine
Philadelphia, Pennsylvania 19104

1837 **LIE, SVERRE O.; LOKEN, AAGOT CHR.; STROMME, JOHAN H.; & AAGENAES, OYSTEIN.** Fatal congenital lactic acidosis in two siblings. I. Clinical and pathological findings. *Acta Paediatrica Scandinavica*, 60(2):129-137, 1971.

Clinical and pathological findings in 2 siblings who died of metabolic acidosis within one week of birth are presented, revealing nearly identical processes in the lungs, liver, thymus, and brains of the two. In the second sibling the acidosis was traced to an accumulation of lactic acid in the serum, but no lactic acid determinations were made in the first infant. An identical condition is suggested by the similarity of pathological findings; the fact that the two infants were siblings suggests a genetic factor. As none of the known conditions causing lactic acidosis were present, patients were considered to have had a primary type of lactic acidosis, probably with biochemical lesion. The cases differed from those previously reported in that: there were no signs of respiratory effort to get rid of the acid load, suggesting damage to the respiratory center; serum potassium levels were elevated in both patients, possibly because of renal failure; there was no aminoaciduria, but a pathological elevation of the protein content of the spinal fluids. There were elevated lactic dehydrogenase and creatine phosphokinase levels in the serum of the second infant. Myelin formation in the brain hemispheres was underdeveloped, suggesting a deficient process in the oligodendroglial cells. Brain pathology in other cases of lactic acidosis reported in the literature is compared with the present cases; it is noted that one case of fructosemia presents a quite similar configuration. (20 refs.) - *N. Jarvis.*

University of Oslo
Rikshospitalet, Oslo, Norway

1838 **GARTLER, STANLEY M.; SCOTT, RONALD C.; GOLDSTEIN, JOSEPH L.; CAMPBELL, BARBARA; & SPARKES, ROBERT.** Lesch-Nyhan syndrome: Rapid detection of heterozygotes by use of hair follicles. *Science*, 172(3983):572-574, 1971.

Direct assay of hypoxanthine guanine phosphoribosyltransferase (HGPRT) activity in single hair follicles from the scalp enables rapid diagnosis of a S who is heterozygous for the X-linked gene responsible for the Lesch-Nyhan syndrome. HGPRT activity in single hair follicles of 4

Lesch-Nyhan heterozygotes, 2 affected male hemizygotes, and 12 normals revealed 3 classes of follicles in the heterozygotes, including those with only deficient activity, those with intermediate activity, and those with only normal activity. Hemizygotes and normals did not show these results. This method may become valuable when it is necessary to make the diagnosis of maternal Lesch-Nyhan heterozygosity during early pregnancy, since it would determine whether or not to offer the patient amniocentesis. (10 refs.) - *B. J. Grylack.*

University of Washington
Department of Genetics
Seattle, Washington 98105

1839 Hyperprolinaemia. *Lancet*, 1(7706):957-958, 1971. (Editorial)

Hyperprolinemia is a rare inborn error of metabolism characterized by raised plasma proline concentrations and increased urinary excretion of proline, hydroxyproline, and glycine. Two types of the disease are distinguished according to enzymatic defect. Type I may be associated with MR, renal abnormalities, hereditary deafness, and EEG abnormalities on photic stimulation. MR and seizures may be present in type II, but deafness and renal lesions have not been described. Hyperprolinemia appears to be transmitted as an autosomal recessive trait. It has been suggested, on the basis of observation, that proline should not be considered an essential element, at least in early life. Thus, the possibility exists that dietary management of infants with the disease may lead to the prevention of some of its deleterious effects. (14 refs.) - *B. J. Grylack.*

1840 **MONCH, E.; STEFAN, H.; & KASER, H.** Neuroblastom Typ Pepper mit Homozystinurie; Beitrag zur Diagnose, Biochemie und Therapie. (Pepper type neuroblastoma with homocystinuria; a contribution to diagnosis, biochemistry and therapy). *Helvetica Paediatrica Acta*, 25(5):530-542, 1970.

Diagnostic studies and laboratory experiments reported here suggest a relationship between vitamin B_6 and the excretion of cystathionine and homocystine. A neuroblastoma metastasizing only to the liver (Pepper type) was diagnosed at the age of 7 weeks in a child now 2½ years old. After irradiation with tele-cobalt rays, vincristine and cyclophosphamide were given alternatively. Initially an increased urinary excretion of vanillylmadelic acid, homovanillic acid, dopamine, 3-methoxytyramine, norepinephrine and free normetanephrine, as well as of cystathionine and homocystine was observed. In the investigation of possible explanations for the cystathionine and the hitherto unreported homocystinuria, intravenous loads of L-methionine and L-serine, as well as oral loads of vitamin B_6 were administered. It was concluded from analysis that vitamin B_6 is an important factor influencing the excretion of cystathionine and homocystine. (29 refs.) - *P. Custers.*

Kinderklinik der Freien Universitat Berlin
Tiefenauspital, Bern, Switzerland

1841 **WELCH, J. PHILIP.** Maternal phenylketonuria. *British Medical Journal*, 1(5739):46-47, 1971. (Letter)

Mass phenylketonuria (PKU) screening in women during pregnancy is impractical. Newborns are already subject to screening and the economics of this has been questioned. This much rarer condition would be difficult to justify economically, and the course of action should one identify a homozygous mother has not been determined. High-risk individuals such as siblings of PKU patients and mothers of MR children have probably already been tested. (6 refs.) - *E. L. Rowan.*

Dalhousie University
Halifax, N.S., Canada

1842 **FARQUHAR, JAMES W.; MILLER, MARION C.; & LINDSAY, GAVIN.** Maternal phenylketonuria. *British Medical Journal*, 1(5739):46, 1971. (Letter)

Two women with phenylketonuria (PKU) have been observed during pregnancy. Neither had received dietary treatment and both had high serum phenylalanine levels and were mildly MR. One could not tolerate the low phenylalanine diet and aborted. The other responded well to dietary management and showed normal serum

phenylalanine levels. She delivered a full-term infant who appeared to be normal in every respect and showed no evidence of PKU. Dietary management appears practical, but women not already known to have PKU would require antenatal screening for identification. (No refs.) - *E. L. Rowan.*

University of Edinburgh
Edinburgh, Scotland

1843 **BEUTLER, E.; KUHL, W.; TRINIDAD, F.; TEPLITZ, R.; & NADLER, H.** β-glucosidase activity in fibroblasts from homozygotes and heterozygotes for Gaucher's disease. *American Journal of Human Genetics*, 23(1):62-66, 1971.

An assay of β-glucosidase activity in skin fibroblast cultures can diagnose Gaucher's disease or its carrier state. The enzyme activity is markedly decreased in the homozygous state and intermediate between homozygous and control values in the heterozygous. An enriched medium produced diminished activity suggesting that fibroblast cultures might lend themselves to experimental manipulation. The assay technique is also valid in amniotic cells and might be used for prenatal diagnosis of Gaucher's disease. (7 refs.) - *E. L. Rowan.*

City of Hope Medical Center
Duarte, California 91010

1844 **OKADA, SHINTARO; VEATH, M. LOIS,; LEROY, JULES; & O'BRIEN, JOHN S.** Ganglioside GM_2 storage diseases: Hexosaminidase deficiencies in cultured fibroblasts. *American Journal of Human Genetics*, 23(1):55-61, 1971.

Skin fibroblast cultures from patients with ganglioside GM_2 storage diseases and controls were found to produce hexosaminidases in the patterns previously reported. The stage of culture did have an effect on enzyme activity. Five children with Tay-Sachs disease showed less than 10% of the control level of hexosaminidase A. A child with Sandhoff's disease had less than 5% of control levels of both hexosaminidase A and hexosaminidase B. A patient with juvenile GM_2 gangliosidosis had 25% control activity of hexosaminidase A. Parents showed values intermediate between patients and controls, thus establishing heterozygosity. This culture technique can be useful in determining enzyme deficiency. (8 refs.) - *E. L. Rowan.*

University of California, San Diego
La Jolla, California 92037

1845 **HANSEN, J.D.L.; FREESEMANN, C.; MOODIE, A. D.; & EVANS, D. E.** What does nutritional growth retardation imply? *Pediatrics*, 47(1):299-312, 1971.

A group of 53 children (South African blacks) who had been hospitalized for kwashiorkor between 6 months and 4 years of age was followed for 10 years to determine physical and intellectual development. Despite the early protein-calorie deficiency 42% of the group reached the normal range (greater than the third percentile) for height and 60% for weight. Forty of these children showed the same intelligence scores as a control group of 40 siblings who had not had kwashiorkor. It appears that given proper treatment and a continuing improved environment, nutritionally deprived children can attain normal physical and mental development. (58 refs.) - *E. L. Rowan.*

Medical School Observatory
University of Cape Town, South Africa

1846 **METCOFF, J.; YOSHIDA, T.; MORALES, M.; ROSADO, A.; URRUSTI, J.; SOSA, A.; YOSHIDA, P.; FRENK, S.; VELASCO, L.; WARD, A.; & ALUBAIDI, Y.** Biomolecular studies of fetal malnutrition in maternal leukocytes. *Pediatrics*, 47(1):180-191, 1971.

The metabolism of both fetal and maternal leukocytes was determined in order to relate it to intrauterine malnutrition (IUM) and growth retardation. Cord blood samples from 16 IUM infants, 24 premature infants, and 33 mature infants were compared with regard to size and enzyme activity. Cells from the IUM group were larger, but the difference did not reach statistical significance. Adenylic kinase and pyruvic kinase activity was diminished in IUM infants and their mothers. A presumptive diagnosis of IUM might

then be made by analyzing maternal leukocytes during pregnancy, but the complex interrelationships of maternal nutrition, leukocyte metabolism, and infant development have yet to be established. (49 refs.) - *E. L. Rowan.*

University of Oklahoma Medical Center
Oklahoma City, Oklahoma 73104

1847 **HILL, LEIGHTON L.; MORRIS, C. RICHARD; & WILLIAMS, ROBERT L.** Role of tissue hypoxia and defective renal acid excretion in the development of acidosis in infantile diarrhea. *Pediatrics,* 47(1):246-253, 1971.

Defective renal excretion of acids secondary to dehydration does not appear to be a major factor in the genesis of the acidosis found in infantile diarrhea. Ten infants hospitalized for diarrhea, dehydration, and metabolic acidosis were followed with serial blood lactic acid and pyruvic acid levels and renal acid excretion measurements. Only 3 infants showed a mild increase in blood lactic acid and renal hydrogen ion excretion was adequate in all. Metabolic acidosis must then have another etiology, and excessive loss of bicarbonate-containing stools is postulated. (25 refs.) - *E. L. Rowan.*

Baylor College of Medicine
Houston, Texas 77025

1848 **GALL, G.; VAITUKAITIS, J.; HADDOW, J. E.; & KLEIN, R.** Erythrocyte Na flux in a patient with Bartter's syndrome. *Journal of Clinical Endocrinology and Metabolism,* 32(4):562-567, 1971.

A 7-year-old boy with Bartter's syndrome manifested by hyponatremia, hypokalemia, hypochloremic alkalosis, dwarfing and MR, was shown to have an increased cell membrane permeability. He was shown to have increased Na active transport, increased Na leak, elevated Na+K dependent ATPase, and increased intracellular sodium in both erythrocyte ghosts and intact red blood cells. The patient showed no response to angiotension infusion and control patients with hyperaldosteronism did not show the same Na flux, suggesting that the basic defect in Bartter's syndrome may be in cell permeability. (14 refs.) - *E. L. Rowan.*

Boston University School of Medicine
Boston, Massachusetts

1849 **PHAROAH, P.O.D.; BUTTFIELD, I. H.; & HETZEL, B. S.** Neurological damage to the fetus resulting from severe iodine deficiency during pregnancy. *Lancet,* 1(7694):308-310, 1971.

Injections of iodized oil were given in a controlled study to women of child-bearing age in the highlands of New Guinea in order to prevent endemic cretinism. This condition is characterized by deaf-mutism, diplegia, squint, and MR. Followup over a 4-year period revealed 26 endemic cretins out of 534 children born to mothers who did not receive oil and only 7 among the 498 who did receive iodine. In at least 6 of these 7 cases the mother was already pregnant at the time of injection. This suggests that neurological damage is done early in pregnancy and that iodine injections before conception could prevent this condition in endemic areas. (16 refs.) - *E. L. Rowan.*

Institute of Human Biology
Goroka, Territory of Papua and New Guinea

1850 **SCRIVER, C. R.; MACKENZIE, S.; CLOW, C. L.; & DELVIN, E.** Thiamine-responsive maple-syrup urine disease. *Lancet,* 1(7694):310-312, 1971.

An infant girl with normal growth but retarded neurological development was found to have the elevated urinary excretion of leucine, isoleucine, and valine characteristic of maple syrup urine disease. Treatment with thiamine, a coenzyme in the breakdown of branched-chain ketoacids, resulted in normalization of the urine. The fact that this child did not require dietary restriction points up the phenotypic variability of this disease and encourages the search for other vitamin responsive hereditary metabolic diseases. (10 refs.) - *E. L. Rowan.*

McGill University
Montreal 108, Canada

1851 **CHASE, H. PETER; DABIERE, CAROL S.; WELCH, N. NOREAN; & O'BRIEN, DONOUGH.** Intra-uterine undernutrition and brain development. *Pediatrics*, 47(3):491-500, 1971.

Guinea pigs undernourished *in utero* showed a reduction in body and brain weight when compared to adequately nourished controls. Fetal wastage occurred in over half the experimental animals. The cerebellum was more affected than the cerebrum, and the decreased weight persisted into adulthood. DNA, protein, cell size, and lipids (cerebroside, sulfatide, and cholesterol) were initially decreased but were generally in the normal range after adequate postnatal nutrition. The increase probably represents glial rather than neuronal elements, however. The guinea pig has 100% of brain cells present at birth and appears analogous to man, who has 75% of cells at birth, for experimental purposes. Good postnatal nutrition appears to overcome much but not all of the damage from intrauterine malnourishment. (27 refs.) - *E. L. Rowan.*

University of Colorado Medical Center
Denver, Colorado 80220

1852 **D'ELIA, R.; & BARONI, A.** Ultrastructure of human skin fibroblasts in gargoylism. *Helvetica Paediatrica Acta*, 25(5):492-496, 1970.

Dermal fibroblasts were obtained by biopsy from 3 boys with mucopolysaccharidosis: one, CA 4½ yrs, with Hunter's syndrome; one, CA 4½ yrs, with Sanfillippo's syndrome; and an 11 month old child with Hurler's syndrome. Electron microscope findings were essentially the same for all 3, showing that the electron-dense vesicular structures filling the cytoplasm were of 2 kinds: one delimited by a smooth membrane formerly interpreted by others as lisosomes, and another delimited by a rough membrane with ribosomes. It is suggested that these structures are enlarged cisternae of the granular and agranular endoplasmic reticulum and of the Golgi complex. (15 refs.) - *P. Custers.*

University of Padua Pediatric Clinic
Padua, Italy

1853 **RAMPINI, S.; ISLER, W.; BAERLOCHER, K.; BISCHOFF, A.; ULRICH, J.; & PLUSS, H. J.** Die Kombination von Metachromatischer Leukodystrophie und Mukopolysaccharidose als selbstandiges Krankheitsbild (Mukosulfatidose) [The combination of metachromatic leukodystrophy and mucopolysaccharidosis as an independent syndrome (Mucosulfatidosis)]. *Helvetica Paediatrica Acta*, 25(5):436-461, 1970.

Three new cases of combined metachromatic leukodystrophy and mucopolysaccharidosis are described. From an analysis and comparison with 3 earlier cases, it is suggested that this combination of disorders represents a rare, clearly defined disease entity with autosomal-recessive transmission. Because of the combination of an anomaly in mucopolysaccharide and sulfatide metabolism the term 'mucosulfatidosis' seems justifiable. The onset of psychomotor retardation is most often at the end of the first or the beginning of the second year. During the third year rapidly progressive deterioration occurs, leading to complete dementia, severe motor deficit, often blindness and deafness, and death between 4 and 12 years of age. The laboratory findings show changes in X-rays, blood picture, urine, cerebrospinal fluid, nerve conduction velocity, and nerve biopsy. EEG findings show mild, if any, changes in the first stages, and marked unspecific alterations in the final stages. (32 refs.) - *P. Custers.*

Kinderspital Zurich, Steinwiesstrasse 75
CH-8032 Zurich, Switzerland

1854 **WHITE, PHILIP L.** The world stops for the hungry. *Journal of the American Medical Association*, 215(1):110-111, 1971. (Editorial)

Until recently it has been politically fashionable to equate malnutrition in childhood with physical and mental retardation. There is no evidence that feeding people makes them smart, but it is indisputable that hunger makes them dull. Evaluation of the influence of early and severe malnutrition on learning ability has been handicapped by the paucity of appropriate means of measurements for infants and young children as well as by the great difficulty in factoring out the other degrees of freedom which influence learning. The dull or unresponsive infant or child may respond to better diet, but if his environment is culturally and economically destitute, he will remain in

intellectual jeopardy. Malnutrition can be involved directly or indirectly in the initiation and perpetuation of a disheartened, listless segment of the population, thus producing considerable drag on society. Malnutrition as it perpetuates a cycle of underachievement should theoretically be easier to ameliorate than other determinants of social interaction, but simple feeding programs should not become substitutes for the loving mother or erode the role of the father. Improving social conditions may be of no benefit if the apathy is related to undernutrition. On the other hand, improving nutritional status may be necessary before social changes can be effective. (No refs.) - *J. C. Moody.*

AMA Council on Foods and Nutrition
Chicago, Illinois

1855 KALLEN, DAVID J. Nutrition and society. *Journal of the American Medical Association*, 215(1):94-100, 1971.

Malnutrition during early development leads to high infant mortality, reduced resistance to infectious disease, delayed physical maturation, and diminished growth. Severe malnutrition may lead to intellectual impairment, but the direct relationship between moderate malnutrition and intelligence is still unknown, because both nutrition and intellectual development are associated with various social factors. The greatest impact of moderate malnutrition on intellectual performance is probably the resulting listlessness and apathy that hinder the child in learning and other situations which relate to life success. While malnutrition is a medical problem, hunger is a social problem, complicated by the fact that the hungry are also subject to various other noxious social conditions. The possible social repercussions of the ways in which we do feed the hungry must be taken into account. For example when children receive their primary sustenance impersonally and outside the family the nurturing role of the mother is seriously undercut, thereby reducing her effectiveness as an agent of socialization. Of greater concern is that the consequence to our affluent society of permitting children to go hungry, while unknown, is crucial to our moral and social well-being. (41 refs.) - *J. C. Moody.*

Michigan State University
East Lansing, Michigan

1856 MILLIAN, STEPHEN J.; KOGON, ALFRED; & KLEIN, SARAH. Rubella antibody levels in commercial gamma globulin preparations. *American Journal of Public Health*, 61(2):353-358, 1971.

Twenty-three lots of commercial gamma globulin (GG) preparations of venous origin and 32 of placental origin were studied for their content of both neutralizing and hemagglutination inhibition antibody to rubella virus. Analysis of results by dates of expiration (from 1962 to 1971) of the materials showed little variation and no clear trend in the neutralizing antibody levels of venous origin, but wide variation with a definite upward trend of neutralizing antibody in placental preparations expiring over the period 1967-1971. This higher antibody content may most plausibly be attributed to differences in donors of the materials used in the GG preparations. Placental extract donors' mean age would probably be considerably younger than donors of venous blood. Since antibody titers to rubella presumably drop with time, GG derived from the older population would tend to be poorer in rubella neutralizing antibody. Furthermore, a larger fraction of rubella susceptibles in the younger group would result in more rubella infections during an outbreak and be reflected in higher rubella antibody contents of GG prepared in post-epidemic years than in pre-epidemic years. Examination of the same preparations for hemagglutination inhibition (HI) antibody revealed little variation and no time trend in titer level. As the HI titers on all samples were quite high, they may perhaps be approaching a biological maximum. (23 refs.) - *J. C. Moody.*

City of New York Department of Health
New York, New York 10016

1857 De MAEGD, M.; & VAN NEVEL, C. Hypothyreoidie tengevolge van een Kongenitaal Erfelijke Stoornis in de Desiodatie van de Iodotyrosines (Hypothyroidism as a consequence of a congenital inherited deficiency in the deiodinase of the iodotyrosines). *Acta Paediatrica Belgica*, 24(2):115-121, 1970.

Four out of 8 siblings, of Moroccan origin, presented with obvious signs of hypothyroidism and goiter. Biological examinations had shown a low iodinase. The fixation of radioactive iodine

was higher than normal, but there was a rapid elimination. After application of marked iodotyrosine, a large part of this was found back in the urine. Chromatography of the plasma after the application of radioactive iodide showed important quantities of marked iodotyrosine in the plasma. From these endocrinological studies, the existence of a congenital inherited deiodinase deficiency was hypothesized. (5 refs.) - *P. Custers.*

Rijksuniversiteit-Gent
De Pintelaan 135
B-9000 Gent, Belgium

1858 CHOW, BACON F.; SIMONSON, MARIA; HANSON, HARLEY M.; & ROEDER, LOIS M. Behavioral measurements in nutritional studies. *Conditional Reflex,* 6(1):36-40, 1971.

In addition to other common indices for measuring dietary adequacy, behavioral measurement can reflect nutritional inadequacy. Studies on the effects of dietary deficiencies have shown marked behavioral impairments in rats deprived of adequate quantities of specific vitamins. In at least 1 study impairment was evident even when body weight did not differ from vitamin-supplemented but pair-fed controls. Recently, experiments have demonstrated that restriction of maternal dietary intake during gestation and lactation results in a variety of physiological and behavioral abnormalities in the progeny. These offspring are stunted, retarded in neuromotor development, impaired in ability to learn a maze, and slow in extinction of the conditioned reflex; they show a low level of exploratory activity, a high degree of emotional behavior, and marked antisocial interaction. In addition, patterns of change with increasing age in some of these measurements also distinguish these experimental animals from controls born of adequately fed dams. (15 refs.) - *S. Glinsky.*

Johns Hopkins University
Baltimore, Maryland

1859 DANIEL, WILLIAM L.; & HIGGINS, JAMES V. Biochemical and genetic investigation of the de Lange syndrome. *American Journal of Diseases of Children,* 121(5):401-405, 1971.

Increased serum α-ketoglutarate and serum glutamate levels may prove significant in the diagnosis of the de Lange syndrome. Ten patients with this syndrome exhibited the anomaly of higher serum α-ketoglutarate and serum glutamate levels. Serum glutaminase, serum glutamic oxaloacetic transaminase (SGOT), serum glutamic pyruvic transaminase (SGPT), and glutamic dehydrogenase activities were within the normal ranges. Hypoaminoaciduria and hypogammaglobulinemia were present in 6 of the patients, but their significance is questionable. Immunological studies indicated normal IgA, IgG, and IgM levels. Family studies revealed elevated or high-normal glutamate levels in the parents and certain siblings of the patients, indicating possible inheritance of hyperglutamicacidemia. All cases appeared to be sporadic with one exception. Synophrys and clinodactyly were noted in relatives of one of the patients. The de Lange syndrome may be the result of an inherited metabolic error, although the exact nature of the enzymatic defect is uncertain. (22 refs.) - *S. Glinsky.*

Illinois State University
Normal, Illinois 61761

1860 PAK, YONG CHA; LIMBECK, GEORGE A.; & KELLEY, VINCENT C. Hypoglycemia in children. *American Journal of Diseases of Children,* 121(1):10-14, 1971.

Retardation of growth in hypoglycemic children may be due to decreased urinary 17-ketosteroid (17-KS). Adrenocortical function was studied in 25 children with hypoglycemia, separated into 2 groups, depending on whether their height was above or below the third percentile. Plasma 17-hydroxicorticosteroid (17-OHCS) concentrations were not significantly different in the 2 groups, and were higher than normal in both. In both groups the urinary 17-OHCS values were quite low considering the high plasma concentrations, but in neither group were they significantly lower than normal. Adrenal responsiveness to adrenocorticotropic hormone (ACTH) was normal as judged by the responses of both plasma and urinary 17-OHCS. In contrast, the control urinary 17-KS levels differed significantly from each other in the 2 groups, those in the group with severe growth retardation being significantly lower than normal and those in the other group being in the low normal range. (18 refs.) - *S. Glinsky.*

University of California, Irvine
Irvine, California

1861 SPERLING, MARK; & DRASH, ALLAN. Evolution of diabetes mellitus from hypoglycemia. *American Journal of Diseases of Children*, 121(1):5-9, 1971.

Two children with idiopathic hypoglycemia of infancy developed diabetes mellitus. One patient, a 16-year-old boy, is insulin dependent, while the other patient, an 11-year-old girl, received tolbutamide. Despite recurrent hypoglycemia, blood glucose values during oral glucose tolerance were consistent with a diagnosis of diabetes, and the immunoreactive insulin responses to a number of stimuli were impaired. Currently accepted glucose-endocrine interrelationships do not explain these observations. Although reports of the progression from hypoglycemia to diabetes are rare, the association appears to be more than fortuitous. An understanding of the evolution of this process may provide valuable insight into the pathophysiology of carbohydrate intolerance, and the role of glucagon requires clearer definition. (27 refs.) - *S. Glinsky.*

University of Pittsburgh School of Medicine
Pittsburgh, Pennsylvania

1862 LOWRY, R. B.; & RENWICK, D.H.G. Relative frequency of the Hurler and Hunter syndromes. *New England Journal of Medicine*, 284(4):221-222, 1971. (Letter)

The relative frequency of the Hunter and Hurler syndromes over a 16 year period in British Columbia is established. Data were taken from the British Columbia Registry for Handicapped Children and Adults for the years 1952-68. The Registry derives its data from discharge diagnoses from all hospitals in the province, children's outpatient clinics, public health units, residential institutions for MRs, physicians' notices of birth, private physicians, and special treatment centers. The incidence rates per 100,000 live births are 1 for Hurler's syndrome, 0.66 for Hunter's syndrome, and 0.33 for Morquio's syndrome. Although the mutation rates for the first 2 syndromes differ, they may in fact be equal. Since determination of these 2 syndromes requires at least a few years, such facilities for long-term collection of data are good sources of frequency of incidence. (3 refs.) - *S. Glinsky.*

University of British Columbia
Vancouver, British Columbia, Canada

1863 SPRANGER, J. W. A new mucopolysaccharidosis. *Journal of Pediatrics*, 78(2):366, 1971. (Letter)

A recent report of a new mucopolysaccharidosis reflects the difficulties of a nosologic classification of these diseases. The patients were thought to have a new mucopolysaccharidosis since they had a normal facial appearance, normal height, and a high amount of heparan sulfate in the urine, along with the features of mucopolysaccharidosis V. None of these findings, however, is inconsistent with Scheie's disease, except for the unusually high heparan sulfate index which may be accounted for by advanced methods of detection. Until further investigations of other patients with mucopolysaccharidosis, utilizing the same advanced techniques, are carried out, a diagnosis of mucopolysaccharidosis V seems to be safe. (9 refs.) - *S. Glinsky.*

University of Wisconsin
Madison, Wisconsin

1864 COLLIPP, P. J.; REZVANI, I.; & BREN, G. A new mucopolysaccharidosis. *Journal of Pediatrics*, 78(2):365-366, 1971. (Letter)

The present inadequacy of traditional criteria for identifying a new mucopolysaccharidosis makes a recently reported new syndrome questionable. Five hundred patients examined over a 1½ year period and selectively screened for mucopolysaccharidosis revealed 16 patients, aged 2 months to 17 years of age, with this disorder. Only 4 patients had clinical and X-ray findings characteristic of the 6 known types of mucopolysaccharidoses, and none resembled cases reported in the literature. Perhaps the best method of identifying a new form of mucopolysaccharidosis is to show that the genetic defect in tissue culture can be corrected by all the known forms of mucopolysaccharidoses except that of the patients of the same form. In the present confused state any proliferation of new syndromes would not seem justified. (7 refs.) - *S. Glinsky.*

Nassau County Medical Center
East Meadow, New York

1865 HORTON, W. A.; & SCHIMKE, R. NEIL. A new mucopolysaccharidosis – Reply.

Journal of Pediatrics, 78(2):366-367, 1971. (Letter)

Patients classified by the authors as having a new form of mucopolysaccharidosis exhibit sufficient differences to cause a renewed rejection of the Scheie's disease diagnosis. The hepatitin sulfate excretion pattern of the patients may change but has remained remarkably consistent over a period of 3 years. Only the development of specific enzyme assays will make identification of new forms of mucopolysaccharidosis possible. The biochemical differences rather than the clinical and genetic similarities should be emphasized so that the presence or absence of heterogeneity can be more clearly delineated. (No refs.) - *S. Glinsky.*

Kansas University Medical Center
Kansas City, Kansas

1866 MAN, EVELYN B.; HOLDEN, RAYMOND H.; & JONES, WALTER S. Thyroid function in human pregnancy. VII. Development and retardation of 4-year-old progeny of euthyroid and of hypothyroxinemic women. *American Journal of Obstetrics and Gynecology*, 109(1):12-19, 1971.

Seventeen children born to hypothyroxinemic women taking adequate thyroid replacement therapy during pregnancy and 27 born to hypothyroxinemic women receiving inadequate thyroid therapy were compared at 4 years of age in regard to development with 192 children born to euthyroid women. Tests of IQ, vocabulary, speech, and fine and gross motor skills were administered to determine relationships between development and retardation of children, on the one hand, and maternal gestational hypothyroidism, on the other. Children of hypothyroxinemic women taking inadequate thyroid replacement therapy scored lower than progeny of euthyroid or hypothyroxinemic women taking adequate therapy. There is an urgent need for thyroxine during the rapid development of the central nervous system *in utero* and during the first 6 to 12 months of a child's extrauterine life. Administration of thyroid replacement therapy to hypothyroxinemic pregnant women was not associated with any adverse effects on the progeny which were recognizable upon developmental examinations of the children. The data suggest that maternal hypothyroxinemia should be corrected during human pregnancies if the development of the child is to be adequate at age 4.(28 refs.) - *B. J. Grylack.*

Rural Route No. 2, Box 370-A
North Stonington, Connecticut 06359

1867 ZAHTZ, HYMAN. Your child and the thyroid gland. Part II. *Jewish Parent*, 22(2):32, 1971.

Hypothyroidism in childhood may begin in the fetal stage or may become an acquired disorder after birth. Congenital hypothyroidism may be a result of genetic abnormalities and maternal exposure to a virus, radioactivity, or certain medications. Acquired hypothyroidism may be due to lack of iodine intake, surgical destruction, autoimmune processes, infectious disease, and disturbance of hormonal balance. Brittle hair, dry and cold skin, large tongue, swollen eyelids, poor appetite, lethargic disposition, constipation, and MR may be signs of hypothyroidism. The disorder can be diagnosed readily by chemical and radiological means. The key to prognosis is early identification, especially in congenital cases. Treatment of hypothyroidism is primarily one of substitution. (No refs.) - *B. J. Grylack.*

No address

1868 POZSONYI, J.; & FRIESEN, H. Growth hormone investigation in patients with mental dysfunction. *Canadian Medical Association Journal*, 104(1):26-29, 1971.

Seventy-three patients between the ages of 6 months and 20 years with either MR or an emotional disturbance accompanied by physical growth disorders were tested for human growth hormone (HGH) abnormalities. Measurements of HGH were aided by stimulation with insulin-induced hypoglycemia and arginine infusion. All but 4 of the 73 patients presented normal HGH responses in the form of a peak value of 7 μg/ml or higher after stimulation. Of the many children with various growth disorders, only those with hypopituitary dwarfism showed evidence of either HGH deficiency or an abnormality in responsiveness of the hypothalamic HGH releasing mechanism to stimulation. Conclusions

could not be drawn as to whether an impaired HGH response following the stimulation test is indicative of a functional impairment of HGH secretion and whether impairment is significant in the growth retardation of patients with mental disorders. (17 refs.) - *B. J. Grylack.*

Children's Psychiatric Research Institute
London, Ontario, Canada

1869 **NAIK, S. N.; & ANDERSON, DAVID E.** Glucose-6-phosphate dehydrogenase deficiency, hemoglobin and haptoglobin types in Mexicans and American Negroes. *Texas Reports on Biology and Medicine*, 29(1):99-107, 1971.

Eighty-seven Mexican and 269 Negro male and 142 Negro female blood donors were screened for glucose-6-phosphate dehydrogenase (G-6-PD) deficiency, hemoglobin variants, and serum haptoglobin types. G-6-PD deficiency was found in 1.1% of Mexican males, 9.3% of Negro males, and 3.8% of 130 Negro females. Abnormal hemoglobins were not observed in the Mexican group. Among Negroes, 21 males and 6 females manifested hemoglobin (Hb) AS for a total frequency of 6.8% and 7 males and 2 females revealed HbAC for a total frequency of 2.3%. An unusual B_2 hemoglobin fraction in addition to the normal A_2 fraction was found in 2 males and 1 female. Three male and 1 female HbAS samples were also G-6-PD deficient. The frequency of haptoglobin (Hp) allele was 0.46±.04 in Mexicans and 0.54±.02 in Negroes. HpO type was not observed in Mexicans but was seen in Negroes along with Hp 2-1M and the 3 common phenotypes. The 2 population groups studied showed striking genetic variations which could indicate differences in their ethnic origin. The finding of G-6-PD deficiency in only 1 out of 87 Mexicans and the absence of abnormal hemoglobins could imply that this group is free from Negro admixture and/or came from areas free from malarial endemicity. The high frequency of G-6-PD deficiency and abnormal hemoglobins in Negro male blood donors has bearing on transfusion and warrants prescreening for these genetic characters. (26 refs.) - *B. J. Grylack.*

Cancer Research Institute
Tata Memorial Centre
Parel, Bombay 12, India

1870 **MORROW, GRANT, III; SCHWARZ, RICHARD H.; HALLOCK, JAMES A.; & BARNESS, LEWIS A.** Prenatal detection of methylmalonic acidemia. *Journal of Pediatrics*, 77(1):120-123, 1970.

The first case of prenatal detection of an inborn error of metabolism by measuring a maternal secretion is reported. Urine collected from a pregnant mother throughout the course of gestation showed 2.6 mg/day excretion of methylmalonate at 20 weeks gestational age. The level increased to 6.7 at 25 weeks, 32.6 at 31 weeks, and 28.6 at 37 weeks. Amniotic fluid obtained at 25, 31, and 39 weeks of gestation also showed the presence of the material. After birth the diagnosis of methylmalonic acidemia was confirmed in the child. Three previous siblings had died at 5 days, 3 weeks, and 4 months of age, respectively, and the third sib was proved to have the vitamin B_{12}-unresponsive form of the disorder. The infant likely was the source of the abnormal product during gestation, since the excretion of the material by the mother ceased immediately after delivery. (18 refs.) - *M. S. Fish.*

3600 Spruce Street
Philadelphia, Pennsylvania 19104

1871 **OKKEN, A.; DE GROOT, C. J.; & HOMMES, F. A.** Nonketotic hyperglycinemia. *Journal of Pediatrics*, 77(1):164-165, 1970. (Letter)

The second documented case of nonketotic hyperglycinemia with severe clinical symptoms during the early neonatal period is reported. The S, a brother of a patient who was MR due to this disorder, had clinical symptoms suggestive of the ketotic form of the disease. He became jerky on the second day and had irregular respiration on the fourth day, was lethargic, and refused his feeding. Exchange transfusion, intubation, and artificial respiration were without effect, and the S died on the sixth day. Examination of urine specimens collected on the second day showed normal excretion of glycine (12 μmole/24 hr/kg body weight); however, the level was 38 μmole/24 hr/kg body weight on the following day. Despite a special dietary regimen, blood levels of glycine rose to pathological values. Biochemical data did not substantiate the diagnosis of ketotic hyperglycemia, which was indicated by the clinical course of the disease. (7 refs.) - *M. S. Fish.*

University of Gronigen School of Medicine
Gronigen, The Netherlands

1872 **LYON, G.; ROSEMBERG, S.; & THIEFFRY, ST.** Une forme rare de mucopolysaccharidose chez l'enfant (A rare form of mucopolysaccharidosis in a child). *Archives Francaises de Pediatrie*, 28(1):83-94, 1971.

The cases of two brothers with a rare form of mucopolysaccharidosis are reported. The disease first became evident at age 3, and it had a slowly progressive course. The characteristics of the disease are essentially: bilateral corneal opacities, claw-hands deformation with stiffness of the limb joints, few radiological abnormalities of the bones, and Gasser's cells in the bone marrow. Intelligence in both cases is normal. Both excrete chondroitin sulfate B and heparitin sulfate in the urine. These cases are compared with similar reports in adults, and their nosology is briefly discussed. The differences between this type of mucopolysaccharidosis and the better known Hurler and Hunter syndromes, particularly with respect to the absence of MR and facial dysmorphia in the 2 cases reported here, are commented upon. (11 refs.) - *P. Custers.*

Hopital Saint-Vincent-de-Paul
Paris 14^{e}, France

1873 **VOLK, B. W.; ADACHI, M.; SCHNECK, L.; & VALENTI, C.** Early evolution of intracytoplasmic inclusion bodies of Tay-Sachs disease. *Journal of Neuropathology and Experimental Neurology*, 30(1):147-148, 1971.

The evolution of intracytoplasmic lipid bodies in Tay-Sachs disease is unknown. Ultrastructural, histochemical, and biochemical studies were performed on the brain and liver of a 21-week-old fetus obtained by therapeutic abortion on a woman who had previously given birth to a child with Tay-Sachs disease. Similar studies were carried out on brain and liver from a 14-week-old infant afflicted with this disease. In both cases there was an absence of hexosaminadase A activity in brain and liver, while increased amounts of G_{M2} ganglioside were found in the brain. Histochemically no visible stored material was demonstrated with various histochemical preparations in the immature neurons of the fetal brain. The infant brain showed fine cytoplasmic vacuoles in the well-developed cerebral neurons which exhibited positive reactions with PAS and LFB stains. Ultrastructurally the fetal brain showed occasional dense bodies in the neuronal cytoplasm consisting of fine granules mixed with a moderately electron-dense amorphous material. Occasionally membrane-bound cytosomes with parallel flat membranes were seen. The endoplasmic reticulum was frequently distended and contained also a granular material which was similar to that in the cytosomes. The neurons of the Tay-Sachs infant contained a small number of intracytoplasmic inclusion bodies of 3 different kinds. (No refs.) - *S. Glinsky.*

No address

1874 **CONSTANTOPOULOS, G.; & DEKABAN, A. S.** Content and distribution of molecular weights of mucopolysaccharides in the brain and other organs of Hurler's patients. *Journal of Neuropathology and Experimental Neurology*, 30(1):144-145, 1971.

Acid mucopolysaccharide (AMPS) molecules in Ss with Hurler's syndrome exhibit increased size in various organs. Fresh and fixed tissues were obtained at autopsy from 2 patients with Hurler's syndrome. Percentage of AMPS content in fresh Hurler tissues per amount of fat-free dry weight was 1.0 for gray matter (normal 0.36), 0.8 for white matter (normal 0.31), 6.5 for spleen, 3.0 for bone, and 1.3 for skin (normal 0.2). AMPS content of fixed tissue was 0.15 for gray matter, 0.15 for white matter, and 0.4 for leptomeninges. A large amount of AMPS was isolated from formalin in which tissue was stored. Heparitin sulfate (HS) was the major AMPS in Hurler liver, while chondroitin sulfate B (CSB) predominated in other tissues. HS from all tissues and part of CSB from the spleen and brain were of small molecular weight. (1 ref.) - *S. Glinsky.*

No address

1875 **KORNFELD, M.** Electron microscopic observations of neurovisceral thesaurismosis in an adult (Kufs type of amaurotic

family idiocy with visceral involvement). *Journal of Neuropathology and Experimental Neurology*, 30(1):144, 1971.

A third case of neurolipidosis accompanied by storage in the viscera is described with attention given to ultrastructural features. The S died at the age of 26 years after 11 years of an illness characterized by cerebellar ataxia, flaccid quadruparesis, and mental impairment. Histologically ballooned neurons were found throughout the central nervous system (CNS) and intestinal plexuses. Electron microscopic examination of the CNS revealed aggregates of lipofuscin bodies, numerous mitochondria and a number of lysosome-like bodies. Lipofuscin bodies and numerous lysosome-like bodies were also found in astrocytes, pericytes and endothelial cells, and large reticulohistiocytes were present in liver, spleen, and bone marrow. No cytoplasmic inclusions characteristic of any storage disease, including various forms of amaurotic family idiocy, were found in either the CNS or viscera. This case seems to be related to a juvenile type of amaurotic family idiocy. (3 refs.) - *S. Glinsky.*

No address

1876 GAMBETTI, P. Accumulation of smooth cisterns, multivesicular bodies and "Zebra" bodies in neurons. A case of peculiar storage condition. *Journal of Neuropathology and Experimental Neurology*, 30(1):148, 1971.

"Zebra" bodies may originate from multivesicular bodies which in turn may form from the smooth cisterns. A cerebral biopsy was performed in a 1½-year-old infant with psychomotor retardation, hypotonia, moderate hepatosplenomegaly and macroglossia. The inspection of the retina indicated pallor of the optic disc. The light microscopic examination was unrevealing. The ultrastructural study showed that neurons contained large amounts of smooth cisterns, vesicles, and multivesicular bodies frequently located in the proximity of the Golgi apparatus. Numerous "Zebra" bodies were present in neuronal perikarya and dendrites. Membranous dense bodies were seen in the astrocytes. Ultrastructural and cytochemical (acid phosphatase) evidence suggests that the "Zebra" bodies may originate from multivesicular bodies which may arise in turn from the smooth cisterns. (No refs.) - *S. Glinsky.*

No address

1877 LEVY, HARVEY L.; MADIGAN, PHYLLIS M.; & PENEVA, PETRANA. Evidence for delayed histidine transamination with histidinemia. *Pediatrics*, 47(1):128-132, 1971.

Maturation of histidine transaminase in early life may be delayed. Free amino acids in urine were analyzed in 7 individuals (age range 6 wks to 8 yrs) with histidinemia. Marked differences in the excretion of imidazolepyruvic acid (IPA), imidazolelactic acid (ILA), and imidazoleacetic acid (IAA) were found between the neonatal and later infancy periods, and the ratios of ILA and IAA to histidine were far greater in later infancy and childhood than in the neonatal period. Concentrations of histidine were greater and concentrations of histidine metabolites were far less during the neonatal period. IPA excretion may be very low or absent in neonates with histidinemia. Therefore, screening processes that detect increased histidine in the urine are more effective in the diagnosis of histidinemia in neonates than procedures that detect only increased excretion of histidine metabolites. (13 refs.) - *J. K. Wyatt.*

Massachusetts General Hospital
Boston, Massachusetts 02114

1878 WALCHNER, ANDREAS M.; KAUFMAN, ROBERT L.; FEIGIN, RALPH D.; & *KAHN, LAWRENCE I. Phenylketonuria. Diagnostic and therapeutic dilemma. *Clinical Pediatrics*, 9(5):263-271, 1970.

Phenylketonuria (PKU) is a primary overflow aminoaciduria and appears to represent a spectrum of disorders rather than a single entity. In classic PKU, there is almost complete inability to hydroxylate phenylalanine to tyrosine, which may be the result of lack of activity of either dihydropteridine reductase or phenylalanine hydroxylase. The system responsible for abnormal brain function in PKU may not be related to the specific genetic effect in a simple manner. Atypical PKU is a partial metabolic block which occurs when genetically-altered phenylalanine hydroxylase converts phenylalanine to tyrosine more slowly than when the enzyme is synthesized by the normal gene but faster than in classic PKU. In classic PKU with delayed maturation of phenylalanine transferase, phenylpyruvic acid may not reach the blood level necessary for

urine spillover until after the blood level of phenylalanine has risen above 50 mg/100 ml. Diagnosis requires examination of all newborns for elevation of phenylalanine levels and differentiation of the cause of phenylalaninemia in cases of positive screening by direct assay of the involved enzyme. Treatment of classic PKU or late PKU begins with diet and frequent monitoring of the blood level of phenylalanine. Present therapy methods are not always successful and may sometimes be difficult to maintain as well as hazardous. (12 refs.) - *J. K. Wyatt.*

*Washington University School of Medicine
St. Louis, Missouri 63110

1879 **McDONALD, T. P.; LOZZIO, CARMEN; & LOTKIN, PAULINE.** Rapid detection and identification of mucopolysaccharides in urine. *Clinical Pediatrics,* 9(5):272-276, 1970.

A screening test for mucopolysaccharides (MPS) reduces the time required for testing and allows for quantitative measurement of urinary MPS without the use of elaborate electrophoretic and chromatographic equipment. This improved alcian blue technique is sufficiently sensitive to establish MPS in normal human urine and was positive in patients with Hurler's syndrome. A paper chromatographic technique provides useful information for differential diagnosis of the mucopolysaccharidoses. The estimate provided is semi-quantitative, but shows 1 to 2-fold increases of MPS fractions over normal urinary levels. These 2 methods are not as quantitative as the Disch carbazole method but they are simple and inexpensive. (9 refs.) - *J. K. Wyatt.*

University of Tennessee
Memorial Research Center and Hospital
Knoxville, Tennessee 37920

1880 **McKUSICK, VICTOR A.** Relative frequency of the Hurler and Hunter syndromes. *New England Journal of Medicine,* 283(26):1467-1468, 1970. (Letter)

This is a reply to criticism of a previous paper on the frequency of Hurler and Hunter syndromes. One critic overlooked the fact that the male with the Hunter syndrome does not reproduce. Two other critics had a common error, the failure to realize that mutation rates are gametic rates. Consanguinity might account for the excess of the Hurler over Hunter syndrome by a ratio of 1.5:1. Also the Hunter syndrome is actually heterogeneous, depending on genes at different loci on the X chromosome. Such problem solving may help to teach biomathematical aspects of population genetics to medical students. (8 refs.) - *E. Kravitz.*

Johns Hopkins University School of Medicine
Baltimore, Maryland

1881 **SCHER, BARRY M.** Relative frequency of the Hurler and Hunter syndromes. *New England Journal of Medicine,* 283(26):1467, 1970. (Letter)

Some of the reasoning by McKusick, in his article on the relative frequency of Hurler and Hunter syndromes, is considered to be erroneous. One basic assumption, that there are equal mutation rates of normal to abnormal genes in both syndromes, is not true biologically. Various other factors are also at play to influence the ratios and birth rates. (1 ref.) - *E. Kravitz.*

Jackson Memorial Hospital
Miami, Florida

1882 **LEVITAN, MAX.** Relative frequency of the Hurler and Hunter syndromes. *New England Journal of Medicine,* 283(26):1466, 1970. (Letter)

A minor mathematical correction is offered to the expected population frequency of Hunter's syndrome discussed in a previous paper by McKusick. The careful wording of the probability expectations is noted favorably. (1 ref.) - *E. Kravitz.*

Mount Sinai School of Medicine
New York, New York

1883 **COLEMAN, MARY.** Serotonin levels in infant hypothyroidism. *Lancet,* 2(7668):365, 1970. (Letter)

High serotonin blood levels were observed in 2 children with hypothyroidism (cretinism) prior to

thyroid therapy. One child had 196 ng/ml as against a normal level of 40-115; the second child had 192 ng/ml, as against a normal level of 100-175. There was increased extremity tone in both cases. Thyroid treatment lowered the serotonin level in one patient from 196 to 148 after 1 week and to 130 after 1 month; tonus improved slightly. Abnormal blood serotonin is related to many diseases associated with mental retardation. Neonatal serotonin screening is under investigation. (7 refs.) - *E. Kravitz.*

Children's Hospital of the
District of Columbia
Washington, D. C. 20009

1884 NADLER, HENRY L.; BIGLEY, ROBERT H.; & HUG, GEORGE. Prenatal detection of Pompe's disease. *Lancet,* 2(7668):369-370, 1970. (Letter)

A case is presented which indicates that the measurement of α-1,4-glucosidase in amniotic fluid supernatant is not a reliable test for the *in utero* detection of Pompe's disease. The use of cultured amniotic cells is recommended as being more meaningful and less subject to error. The patient tested had previously borne a child with Pompe's disease. (5 refs.) - *E. Kravitz.*

Northwestern University Medical School
Chicago, Illinois

1885 BEARDMORE, THOMAS D.; FOX, IRVING H.; & KELLEY, WILLIAM N. Effect of allopurinol on pyrimidine metabolism in the Lesch-Nyhan syndrome. *Lancet,* 2(7677):830-831, 1970. (Letter)

The effects of allopurinol on the urinary excretion patterns of purines and pyrimidines in 2 children with Lesch-Nyhan syndrome were studied. Allopurinol (200-300 mg/day) decreased uric acid excretion and increased equimolar hypoxanthine and xanthine excretion; orotidine excretion increased, but there was no orotic aciduria. Explanations include the possibility that neither allopurinol ribonucleotide nor xanthosine-5′-monophosphate formation is necessary for the inhibitory effect of allopurinol on pyrimidine biosynthesis. (4 refs.) - *E. Kravitz.*

Duke University Medical Center
Durham, North Carolina

1886 KANG, ELLEN SONG; SOLLEE, NATALIE D.; & GERALD, PARK S. Results of treatment and termination of the diet in phenylketonuria (PKU). *Pediatrics,* 46(6):881-890, 1970.

In an investigation of the value of a low phenylalanine diet in the treatment of phenylketonuria (PKU), the clinical course of 82 PKU patients was reviewed, with special attention paid to the level of intellectual functioning. Most children had multiple evaluations over a cumulative period of up to 7 years. IQ findings indicated that those patients diagnosed and treated before 3 weeks, as well as atypical PKU patients, have modal IQs within the normal range. Nineteen children treated between 8 and 42 months did not differ in mean IQ from the 11 children who received no treatment. Significant correlation between average IQ and age at initiation of diet was found for the entire group of treated PKU patients. In comparison to unaffected siblings, patients treated before 3 weeks showed favorable intellectual development, whereas those treated between 3 and 6 weeks performed slightly less well than their siblings on IQ testing. Deterioration was not observed in 26 patients after dietary therapy had been discontinued. A predominance of males was found in the atypical PKU group. (33 refs.) - *B. J. Grylack.*

Children's Hospital Medical Center
Boston, Massachusetts 02115

1887 BOLEY, SCOTT J.; COHEN, MICHAEL I.; & GLIEDMAN, MARVIN L. Surgical therapy of glycogen storage disease. *Pediatrics,* 46(6):929-933, 1970.

Early surgical intervention should be used in the treatment of selected patients with glycogen storage disease due to evidence that other treatment modalities have frequently been unsuccessful in preventing permanent psychomotor retardation secondary to repeated hypoglycemic episodes. A 5-year-old Caucasian female was found to have made favorable gains from a transabdominal end-to-side portacaval shunt. During limited experience with surgical therapy

for glycogen storage disease, it has been observed that the total portal venous diversion has produced a marked relief in hypoglycemia, hyperlipidemia, hyperuricemia, and acidosis, and that there has been no apparent deglycogenation of the liver. Since the primary value of this operation consists in the prevention of hypoglycemia and associated acidosis, it is indicated only in children in whom these are major problems. Early shunting may prevent the development of severe hepatomegaly and related respiratory difficulties as well as cerebral damage due to episodic hypoglycemia. (17 refs.) - *B. J. Grylack.*

Albert Einstein College of Medicine
Bronx, New York 10467

1888 **CUTTINO, JOHN T., JR.; SUMMER, GEORGE K.; HILL, HOYLE D., JR.; & MITCHELL, B. JEAN.** Response to medium chain triglycerides in von Gierke's disease. *Pediatrics*, 46(6):925-929, 1970.

The effects of ingested carbohydrate, when the fat in the diet was provided by long chain and medium chain triglycerides or serum lipid levels, were studied in a patient with von Gierke's disease who exhibited hypoglycemia, hepatomegaly, marked hyperlipemia, and eruptive xanthomas. Increased carbohydrate in the diet resulted in increased lipemia, represented mainly by the triglyceride fraction. Replacement of medium chain triglycerides for long chain triglycerides in the diet when carbohydrate consumption was at normal levels resulted in significantly decreased lipemia, disappearance of eruptive xanthomas, and reduction in liver size. Although the hyperlipemia appears to be carbohydrate-induced in this patient, full clarification of the mechanisms involved in production of the high lipid levels in von Gierke's disease still requires further study. Nevertheless, it can be recommended that excess carbohydrate be avoided in the diet and that substitution of medium for long chain triglycerides be utilized as the sensible treatment regimen. (12 refs.) - *B. J. Grylack.*

University of North Carolina School of Medicine
Chapel Hill, North Carolina 27514

1889 **OH, WILLIAM; D'AMODIO, MABEL D.; YAP, LUCY L.; & HOHENAUER, LEONHARD.** Carbohydrate metabolism in experimental intrauterine growth retardation in rats. *American Journal of Obstetrics and Gynecology*, 108(3):415-421, 1970.

In an investigation of carbohydrate metabolism during experimental intrauterine growth retardation in rats, unilateral uterine artery ligation was performed at 17 days of gestation in 34 pregnant rats to provide an animal model which would allow evaluation of fetal hepatic glycogen reserve *in utero*, fetal blood glucose values at birth, and effects of maternal glucose infusion, prior to delivery of the fetus, on the fetal glycogen reserves and glucose homeostasis at birth. One hundred and fifty-eight control and 141 intrauterine growth retarded fetuses delivered at 21 days of gestation showed that major organ weights were smaller in the abnormal fetuses and that hepatic glycogen contents were lower in the abnormal fetuses during the last 2 days of pregnancy. Intravenous glucose infusion to the maternal rats 2 hours prior to delivery raised the fetal blood glucose to a hyperglycemic range and replenished the previously observed lower hepatic glycogen in the abnormal rats. It is possible that if intrauterine growth retardation is predicted during labor, intravenous glucose infusion to the mother in labor may improve the carbohydrate reserve of the infant even before birth and during the first hours after birth. (16 refs.) - *B. J. Grylack.*

Harbor General Hospital
Torrance, California 90509

1890 **LUSE, SARAH A.; RHYS, ANN; & LESSEY, ROBERT.** Effects of maternal phenylketonuria on the rat fetus. *American Journal of Obstetrics and Gynecology*, 108(3):387-390, 1970.

In an investigation of the deleterious effects of maternal phenylketonuria on heterozygote offspring in the human being, fetuses spontaneously delivered from experimentally induced phenylketonuric rats whose gestation was lengthened by 1 to 3 days were examined. Delayed intrauterine development despite prolonged gestation and fetal resorption were observed, and bilateral cataracts were found in 7 out of 24 fetuses. Excessive abortions, congenital malformations, and MR are associated with a high phenylalanine level

during pregnancy. The high incidence of these problems in those infants born to phenylketonuric mothers but nonhomozygous for phenylketonuria indicates that a high maternal phenylalanine level is detrimental to the fetus. Human and animal fetal phenylalanine levels have been shown to be from 2 to 6 times maternal plasma levels. Placental active transport of phenylalanine is a threat to the infant not only because of the high concentration of phenylalanine in fetal blood at a time when the blood brain barrier is incompletely developed but also because of amino acid imbalance, which may lead to deficiency of certain essential amino acids through competitive absorption of phenylalanine. (11 refs.) - *B. J. Grylack.*

630 West 168th Street
New York, New York 10032

1891 QUINLAN, C. D.; & MARTIN, E. A. Refsum's syndrome: Report of three cases. *Journal of Neurology, Neurosurgery and Psychiatry*, 33(6):817-823, 1970.

Investigation of 3 cases of Refsum's syndrome, all of whom were siblings from the same family, revealed the cardinal diagnostic signs of chronic polyneuritis, cerebellar ataxia, atypical retinitis pigmentosa, and an increased protein content of cerebrospinal fluid without pleocytosis. Evidence was provided that high accumulation of phytanic acid is the cause of the disease and that a correlation exists between the degree of clinical disability and the amount of phytanic acid in the serum. Although 1 patient appeared to benefit from institution of a low phytol and phytanic acid diet, the effectiveness of dietary treatment is still not established. Further trials of the dietary treatment regimen would prove worthwhile. (25 refs.) - *B. J. Grylack.*

Adelaide Hospital
Dublin, Ireland

1892 JEWELEWICZ, RAPHAEL; & NACHTIGALL, LILA E. Pseudopseudohypoparathyroidism and pregnancy: A case report. *Obstetrics and Gynecology*, 37(3):396-401, 1971.

Although individuals with pseudopseudohypoparathyroidism are less fertile, normal pregnancy and delivery is possible. A 20-year-old MR woman (WAIS IQ 47) presented the typical congenital anomalies of the pseudopseudohypoparathyroidism syndrome and had normal calcium and phosphorus values. Although she conceived and bore a normal child, the prognosis for conception and normal delivery was guarded because of short stature, menstrual irregularities, and a small, narrow, bony pelvis. (17 refs.) - *J. K. Wyatt.*

Columbia University College of
Physicians and Surgeons
New York, New York 10032

1893 McDONALD, JOHN A.; & KELLEY, WILLIAM N. Lesch-Nyhan syndrome: Altered kinetic properties of mutant enzyme. *Science*, 171(3972):689-691, 1971.

A hypoxanthine-guanine phosphoribosyltransferase (PRT) enzyme was found in a male (CA 10 yrs) with Lesch-Nyhan syndrome, providing direct evidence for genetic heterogeneity within a group of patients with this syndrome. When an assay with a very high concentration of magnesium 5-phosphoribosyl-1-pyrophosphate (MgPRPP) was used, activities of the PRT enzyme ranged from 8 to 34% of normal in erythrocytes. The mutant enzyme also displayed sigmoidal kinetics with this substrate and an increased Michaelis constant for guanine and hypoxanthine. It is unusual to detect a human genetic disorder which is associated with altered kinetic constants. The sigmoidicity of the MgPRPP rate curve observed with the mutant enzyme may be due to cooperative interaction of a substrate with an enzyme or allosterism. (10 refs.) - *J. K. Wyatt.*

Duke University Medical Center
Durham, North Carolina

1894 PENFOLD, J. L.; KNEEBONE, G. M.; WELLBY, M.; & OLDFIELD, R. K. Growth retardation and thyroxine-binding globulin deficiency. *Archives of Disease in Childhood*, 46(245):115-117, 1971.

A second case of thyroxine-binding globulin (TGB) deficiency displayed delayed bone age and growth retardation and differed from the first

case in that TGB levels were low but not zero. The 9-year-old male was short of stature (114 cm) and exhibited a loss of visual acuity in his right eye. Bone age was between 6 and 6½ years. Total T_4 iodine was low but was not supported by low levels of T_3 uptake, free T_4 index, or ^{132}I uptake. Investigation of T_4 binding in the serum revealed normal thyroxine binding prealbumin and strikingly depressed TGB (5mg/ml serum). No cause was found for the growth retardation. Consistent low levels of TGB suggest a genetic deficiency. Mode of inheritance may be by spontaneous mutation. It is important to assess other parameters of thyroid function, such as the T_3 resin uptake, before prescribing thyroid replacement therapy for children with low total thyroxine iodine. (10 refs.) - *J. K. Wyatt.*

Hospital for Sick Children
Toronto, Ontario, Canada

1895 **YU, J. S.; O'HALLORAN, M. T.; & STUCKEY, S. J.** False negative screening tests in phenylketonuria. *Archives of Disease in Childhood*, 46(245):124-125, 1971.

A false negative finding on a screening test for PKU can result if the infant is tested too early in life, is feeding poorly, or is vomiting. In the 2 cases described, diagnosis of PKU was not made at initial testing because the infants were not feeding well and were vomiting. There should be an adequate protein intake for 4 days prior to routine screening tests. (3 refs.) - *J. K. Wyatt.*

Royal Alexandria Hospital for Children
Camperdown NSW, Australia

1896 **HARRIES, J. T.; PIESOWICZ, A. T.; SEAKINS, J.W.T.; FRANCIS, D.E.M.; & WOLFF, O. H.** Low proline diet in Type I hyperprolinaemia. *Archives of Disease in Childhood*, 46(245):72-81, 1971.

Proline appears to be a "semi-essential" amino acid in the developing infant. A 7-month-old male presented hypocalcemic convulsions and malabsorption and was diagnosed as Type I hyperprolinemia. He had grossly raised plasma levels of proline as well as increased urinary excretion of proline, hydroxyproline, and glycine. There was evidence of delayed neurological development and associated abnormalities of the bones, renal tract, and electroencephalogram. Dietary proline was restricted at 9 months, resulting in reduction of plasma levels of proline to normal. A low proline diet was continued until age 27 months when additional detailed studies evidenced persistence of the biochemical abnormality. After dietary treatment was initiated, mental development improved, physical growth was satisfactory, and abnormalities related to electroencephalogram, and renal, skeletal, and intestinal areas disappeared. (20 refs.) - *J. K. Wyatt.*

Hospital for Sick Children
London, England

1897 **NEVILLE, B.G.R.; HARRIS, RUTH F.; STERN, DIANA J.; & STERN, J.** Maternal histidinaemia. *Archives of Disease in Childhood*, 46(245):119-121, 1971.

An apparently normal male infant was born to a woman (CA 23, WAIS IQ 100) with histidinemia. The mother had grand mal epilepsy. Two urine samples revealed an excess of imidazole pyruvic acid, and her blood histidine was more than 3 times normal (7, 8, and 10 mg/100 ml) on 3 occasions. The histidine load test was characteristic of histidinemia. Her mother and 2 siblings also had histidinemia. When examined at age 4½ years, the boy exhibited normal physical and mental (Stanford-Binet IQ 107) development. Cardiovascular, respiratory, and alimentary systems were normal, as were EEG findings. Serum histidine level was well within the normal range, and excessive imidazole derivatives were not detected in the urine. "Phenistix" and ferric chloride tests produced negative results. It may not be necessary to treat a histidinemic mother with a blood histidine below 10 mg/100 ml. (8 refs.) - *J. K. Wyatt.*

National Hospital
London, WC1, England

1898 **DOBBING, J.** Undernutrition and the developing brain: The use of animal models to elucidate the human problem. In: Stoelinga, G.B.A.; & Van Der Werff

Ten Bosch, J. J., eds. *Normal and Abnormal Development of Brain and Behaviour.* Baltimore, The Williams & Wilkins Co., 1971, p. 20-30.

In many children, irreversible mental deficits stem from malnutrition-induced growth restriction. Even in the most privileged homes there may be diminished potential associated with low birth weight. Long-term effects of poor nutrition on the developing brain depend on the time relationship of the undernutrition with the brain's growth spurt—a period covering the last half to one-third of gestation and the first 18 months to 2 years of postnatal life. This demonstrates the likelihood of brain deterioration during late pregnancy when malnutrition exists and the importance of good nutrition in the early postnatal period. High nutrition in these periods can accelerate brain development. Results of animal experimentation—which can be extrapolated to humans—have revealed the nature of cerebellar growth and the vulnerability of the cerebellum to growth retardation. (26 refs.) - *B. Berman.*

1899 WIDDOWSON, E. M. Effects of early malnutrition on general development in animals. In: Stoelinga, G.B.A.; & Van Der Werff Ten Bosch, J. J., eds. *Normal and Abnormal Development of Brain and Behaviour.* Baltimore, The Williams & Wilkins Co., 1971, p. 39-49.

Undernutrition, whether before or after birth, hinders growth, although all parts of the body are not retarded equally. The brain, when food is short, is in a favored position; undernourished rats, given unlimited food from weaning at 3 weeks, failed to reach the weight gain of well-nourished ones, but the brain showed best results. Initially undernourished pigs made rapid gains when given abundant food, but, at maturity, all organs were smaller than in normal animals. Complete recovery, even of the brain, was not possible, and no organ showed as much DNA as in the corresponding organ of the normal ones. Skeletal development, like that of soft tissues, can also be hindered by malnutrition before or soon after birth. (7 refs.) - *B. Berman.*

1900 VAN GEMUND, J. J.; & DE ANGULO, M. S. LAURENT. The effects of early hypothyroidism on IQ, school performance, and electroencephalogram pattern in children. In: Stoelinga, G.B.A.; & Van Der Werff Ten Bosch, J. J., eds. *Normal and Abnormal Development of Brain and Behaviour.* Baltimore, The Williams & Wilkins Co., 1971, p. 299-313.

Congenital hypothyroidism provides a biological model for studying brain-thyroid relationships in CNS development. A survey of all children with hypothyroidism (46 with a congenital type, 18 with an acquired type) in the Leiden, Holland, Department of Pediatrics in 1952 showed an IQ of less than 90 in 28% of the congenitals and in 56% of the acquired type. For the congenitals, a mean IQ of 71 was attained when thyroxine substitution was started before age 10 months, and an IQ of 83 when started later. Only 31% of the congenitals were found ready for schooling at age 6 years, with only 38% achieving normal progress through primary school. Characteristic learning disabilities included high distractibility, short attention span, and very poor arithmetic performance (this pattern was not seen in the acquired type). Thirty percent needed speech therapy and 50% had motor-coordination problems. EEG studies showed disturbed electrical brain activity. Familial studies showed that, so far as social and genetic factors were involved, there would have been average intellectual development if there had been no thyroid deficiency. (46 refs.) - *B. Berman.*

1901 BERMAN, JULIAN L. Phenylketonuria. *American Family Physician,* 3(2):113-120, 1971.

This survey of PKU in a largely Caucasian population has found that patients receiving early dietary treatment to limit phenylalanine intake eventually, despite some learning defects, score in the 80-90 IQ range. The survey considered clinical, therapeutic, diagnostic, metabolic, and prognostic aspects, while analyzing evaluative criteria and the findings of the National Collaborative Program for the Study of PKU, and defined several criteria for diagnosing PKU: (1) Blood phenylalanine levels greater than 20 mg percent on more than one occasion during the first 121 days of life; (2) persistence of these levels when challenged with evaporated milk at 3 months of age; (3) urine showing the presence of phenylketones during the newborn and challenge

periods; and (4) normal blood tyrosine levels. Of 7 cases demonstrating some of the problems associated with high blood-phenylalanine levels, 3 illustrated PKU and 4 hyperphenylalaninemia, the latter possibly signifying transient PKU, phenylalanine transaminase deficiency, or a transient or permanent hyperphenylalaninemic variant. The survey warns of laboratory errors in determining blood-phenylalanine levels (the optimal range is as yet undetermined). (No refs.) - *B. Berman.*

Chicago Medical School
Chicago, Illinois

1902 LOTT, IRA T.; WHEELDEN, JULIE A.; & LEVY, HARVEY L. Speech and histidinemia: Methodology and evaluation of four cases. *Developmental Medicine and Child Neurology*, 12(5):596-603, 1970.

The speech and language abnormalities noted in histidinemia seem not to be due to a focal lesion caused by metabolic disorder. Four unrelated children with histidinemia or histidase deficiency were studied specifically for characteristics of speech and language. Each child received 5 speech tests, psychometric testing, and a hearing test. Two of the children with normal intelligence were also normal in all aspects of speech and language. The third child, who exhibited subnormal intelligence and emotional immaturity, had normal speech but manifested poor use of syntax on language evaluation. The fourth child, who was mentally retarded and had a moderate hearing loss, also had normal speech but had poor auditory and speech discrimination as well as poor digit recall on language testing. Speech discrimination in this last child became normal when the intensity of the auditory stimulus was increased. A review of the literature indicates that most histidinemic children with speech or language abnormalities also have normal intelligence, while most histidinemic children with normal intelligence also had normal speech and language. Thus these abnormalities noted in histidinemia are not due to a focal lesion caused by metabolic disorder, but may be related to cultural deprivation, mental subnormality, or hearing loss. (33 refs.) - *S. Glinsky.*

Harvard Medical and Massachusetts
General Hospital
Boston, Massachusetts

1903 OLDENDORF, WILLIAM H.; SISSON, WARDEN B.; MEHTA, ARVIND C.; & TRECIOKAS, LEO. Brain uptake of selenomethionine Se 75. *Archives of Neurology*, 24(5):423-430, 1971.

The γ emissions of selenomethionine Se 75 may be useful in clinical studies of blood brain barrier (BBB) penetration by amino acids which share the same carrier mechanism facilitating BBB passage of selenomethionine Se 75. The uptake of selenomethionine Se 75 after intravenous injection in rat was studied in the brain, pancreas, plasma, muscle, scalp, and skull. The data indicated that selenomethionine Se 75 enters rat brain from blood via the same rate-limiting mechanism as methionine and phenylalanine. Acute preloading with intravenously administered nonradioactive L-methionine or L-phenylalanine resulted in a reduction of brain uptake of selenomethionine Se 75. Phenylalanine is a more effective inhibitor than methionine. Measurable inhibition of brain uptake of selenomethionine Se 75 was demonstrated at lower plasma levels of phenylalanine than commonly found in clinical phenylketonuria. Ventriculo-cisternal perfusion studies in rabbits demonstrated bidirectionality of carrier mediated exchange of selenomethionine Se 75 between blood and brain. (22 refs.) - *S. Glinsky.*

Wadsworth Hospital, Veterans
Administration Center
Los Angeles, California 90073

1904 CALLIS, L.; CASTELLO, F.; GORTUNY, G.; VALLO, A.; & BALLABRIGA, A. Effect of hydrochlorothiazide on rickets and on renal tubular acidosis in two patients with cystinosis. *Helvetica Paediatrica Acta*, 25(6):602-619, 1970.

The effect of hydrochlorothiazide in treating two cases of cystinosis with severe growth retardation and vitamin D resistant rickets in children is explained and examined. The first patient, whose hyperchloremic acidosis was of proximal tubular origin, was treated with a 6g/day dose of Na-bicarbonate per os until a HCO_3 blood level of 20-22 mEq/1 was reached. Two months later there was no improvement of the rachitic lesions. The following treatment was then instituted in both cases: hydrochlorothiazide 1.5-2 mg/kg per day; potassium bicarbonate 20 mEq per day.

Normal food intake. After 5 days the plasma standard bicarbonate, the plasma pH, and plasma Cl became normal in the first patient; the urinary Ca excretion was between 3.7 and 7 mg/kg per day, and urinary P excretion decreased to a quasi normal level in both cases. At the same time there was a marked elevation of the RP percentage. In the first patient the threshold and Tm bicarbonate ranged quasi normal level. After 1 month of this treatment the bone lesions almost disappeared. Vitamin D was not given to these children during this time. Whenever this treatment was discontinued there was a relapse of the plasma and urinary alterations. After one year of treatment with hydrochlorothiazide, there was complete correction of the acidosis in the first patient, and the normalization of the Ca and P urinary excretion as well as recalcification of the rachitic bones persisted in both cases. (13 refs.) - *J. C. Moody.*

Children's Hospital of the "Seguridad Social"
Barcelona, Spain

1905 O'BRIEN, JOHN S. Current concepts: Ganglioside-storage diseases. *New England Journal of Medicine*, 284(16):893-898, 1971.

Clinical features, genetics, chemistry and enzyme defects, and detection of affected patients and carriers, including prenatal diagnosis, are described for the 5 recognized ganglioside-storage diseases (Tay-Sachs disease, Sandhoff's disease, juvenile GM_2 gangliosidosis, generalized gangliosidosis, and juvenile GM_1 gangliosidosis) all of which involve progressive mental and motor deterioration, with onset in childhood and ultimate fatal outcome; autosomal recessive inheritance; neuoronal lipidosis secondary to storage of a specific ganglioside; storage of structurally related glycolipids, polysaccharides, or glycoproteins; and absence or severe deficiency of specific lysosomal hydrolase enzymes. No treatment is available for these diseases other than palliative therapy. The most effective approach to their control is the detection of heterozygous carriers followed by family planning and the prenatal diagnosis of affected fetuses followed by selective abortion. (9 refs.) - *J. C. Moody.*

University of California, San Diego
La Jolla, California 92037

1906 DUBOIS, REUBEN S.; RODGERSON, DENIS O.; SLOVIS, THOMAS L.; HAMBIDGE, K. MICHAEL; & BIANCHI, THOMAS A. Triethylene tetramine dihydrochloride in Wilson's disease. *Lancet*, 2(7676):775, 1970. (Letter)

Triethylene tetramine dihydrochloride (T.E.T.A.) has a place in the treatment of Wilson's disease patients in whom D-penicillamine produces a toxic reaction. The patient was a 13-year-old boy who had a 2-year history of liver disease. After confirmation of the diagnosis of Wilson's disease, D-penicillamine treatment was started at a dose of 1g per day. The patient developed a striking cupriuresis of 2387μg per 24 hours while his white cell count fell rapidly to below 2,500 per c mm. Lower doses resulted in similar reactions, and subsequently the patient was given T.E.T.A. at a dose of 1g per day. Urinary copper excretion rose to 1,334 μg per 24 hours, and the white cell count remained stable at 4,000 per c mm. No other signs of toxicity developed, and after 6 weeks of T.E.T.A. treatment the patient was recommenced on D-penicillamine treatment with no adverse effects. T.E.T.A. is therefore of value in Wilson's disease treatment, although D-penicillamine remains the agent of choice. (1 ref.) - *S. Glinsky.*

University of Colorado Medical Center
Denver, Colorado

1907 YATSU, FRANK M. Sphingolipidoses. *California Medicine*, 114(4):1-6, 1971.

The infantile form of Niemann-Pick disease accounts for 85% of the cases of sphingolipidoses. This disease is characterized by psychomotor retardation, mental deterioration, seizures, and spasticity. Lipid analysis reveals an absence of sphingomyelinase and an excess of sphingomyelin. Both Krabbe's and Gaucher's disease have similar symptoms to Niemann-Pick disease. In Krabbe's disease, there is diffuse demyelination of white matter in the brain and an accumulation of globoid cells which exhibit a high concentration of galactocerebroside. Sulfotransferase is the suspected enzyme deficiency. In Gaucher's disease, there is an absence of β-d-glucosidase and a glucocerebroside lipid accumulation. Characteristic symptoms of metachromatic leukodystrophy are in the motor area. The central and peripheral nervous systems and some

organs have intracellular lipids, and there are deposits of sulfatide within the white matter cells as well as demyelination of white matter. Tay-Sachs disease generally occurs in Jewish children. It develops during the first year of life and death usually occurs after 2 to 4 years. Symptoms include MR and myoclonic seizures. Membranous lamellated bodies which reveal an increase of monosialoganglioside are present. Diagnosis prior to birth may be possible when a deficiency of hexosaminidase is found. (31 refs.) - *J. K. Wyatt.*

San Francisco General Hospital
San Francisco, California 94110

1908 **PADEH, BARUCH; & NAVON, RUTH.** Diagnosis of Tay-Sachs disease by hexosaminidase activity in leukocytes and amniotic fluid cells. *Israel Journal of Medical Sciences,* 7(2):259-263, 1971.

Determination of hexosaminidase in amniotic fluid cells permits antenatal detection of fetuses of heterozygous pregnant women affected with Tay-Sachs disease. Hexosaminidase activity was determined fluorometrically in peripheral leukocytes from 12 normal subjects, 13 obligatory heterozygotes, and 6 children with Tay-Sachs disease and in cultured and uncultured amniotic fluid cells from 7 pregnant women. There was a complete deficiency of hexosaminidase A in leukocytes from children with Tay-Sachs disease. Parents of children with Tay-Sachs disease exhibited an intermediate level of hexosaminidase A when compared with normal subjects. Level of hexosaminidase A activity can be used to distinguish individuals who are heterozygous for Tay-Sachs disease. (10 refs.) - *J. K. Wyatt.*

Government Hospital
Tel-Hashomer, Israel

1909 **CRAVIOTO, JOAQUIN.** Nutrition and learning in children. In: Springer, Ninfa Saturnino, ed. *Proceedings of a Conference on Nutrition and Mental Retardation.* Ann Arbor, Michigan, University of Michigan Publications Distribution Service, 1971, p. 25-44.

Malnutrition in early life may obstruct intellectual functioning and contribute to learning disabilities later in life. Children who survive a severe episode of chronic malnutrition that interfered with adequate development of reading and writing are at higher risk of essential intellectual deficits and socioeconomic inadequacies. Adequate nutrition is crucial for optimal growth; in mental development food and feeding display 3 complementary dimensions: (1) physiological, (2) psychological, and (3) psychosocial. All severely malnourished children admitted to the nutrition ward of the Hospital Infantil in Mexico in 1959 showed below-age-norm scores in all behavior fields. As recovery occurred, developmental quotients increased in most Ss. Malnutrition may indirectly affect CNS function through loss of learning time, interference during critical learning periods, and changes in motivation and personality. (No refs.) - *B. Berman.*

Hospital Infantil de Mexico
Mexico City, Mexico

1910 **WAISMAN, HARRY A.; & BROWN, ELEANOR S.** Dietary intervention in maple syrup urine disease and phenylketonuria. In: Springer, Ninfa Saturnino, ed. *Proceedings of a Conference on Nutrition and Mental Retardation.* Ann Arbor, Michigan, University of Michigan Publications Distribution Service, 1971, p. 44-74.

The familial nature of most MR cases has become clear through study of inborn errors of metabolism, in which there is a disorder in a metabolic process that disturbs or blocks orderly metabolic progression. The block causes a build-up of some unmetabolized substance (e.g., carbohydrates) in the brain, with consequent retardation effects. Knowledge of genetics, pediatrics, and biochemistry can help prevent MR by early recognition of a problem and institution of therapy. Maple syrup urine disease, a rarer error than phenylketonuria (PKU), is rather easy to treat if most of the leucine, isoleucine, and valine is removed from the diet. Adding milk provides some natural protein. In PKU, metabolization of phenylalanine to tyrosine is blocked by absence of an enzyme; build-up of phenylalanine in the urine is detected by a simple test. Early dietary treatment has been effective in many cases. Retardation can be prevented frequently by early diagnosis and dietary treatment. (16 refs.) - *B. Berman.*

University of Wisconsin Medical Center
Madison, Wisconsin

MEDICAL ASPECTS – Etiologic Groupings

New Growths

1911 **HAICKEN, BARRY N.** Neurofibromatosis in childhood. *Journal of Pediatrics,* 77(5):923, 1970.

The use of the term "hemi hypertrophy" in a discussion of neurofibromatosis in childhood was unclear. The term may refer to bony overgrowth with localized hypertrophy of adjacent tissues or to congenital overgrowth of one-half or one side of the body. The latter is exceedingly rare and also associated with hamartomas; if noted, it merits further investigation. (1 ref.) - *E. L. Rowan.*

Jacobi Hospital
Bronx, New York 14620

1912 **WILKINS, ROBERT H.; & BRODY, IRWIN A.** Von Recklinghausen's neurofibromatosis. *Archives of Neurology,* 24(4):374-377, 1971.

A case of a 55-year-old female with neurofibromatosis over almost the entire epidermis is presented, which was described by von Recklinghausen in 1879. Von Recklinghausen noted the association of the multiple skin tumors with the peripheral and autonomic nerves. Evidence for this association of neuromas and cutaneous fibromas was the nature of the neoplastic tissue in both tumor types, the penetration of the undersurface of skin tumors by nerve tumors, and the presentation of an arrangement in the cutaneous fibromas different from the usual multiple fibromatous neoplasms. Present research has documented neoplasms and malformations involving the brain, spinal cord, meninges, autonomic ganglia, and the roots of the cranial and spinal nerves, as well as anomalies in other tissues. (9 refs.) - *S. Glinsky.*

Duke University Medical Center
Durham, North Carolina 27706

1913 **LEUMANN, ERNST P.; BAUER, ROBERT P.; SLATON, PAUL E.; BIGLIERI, EDWARD G.; & *HOLLIDAY, MALCOLM A.** Renovascular hypertension in children. *Pediatrics,* 46(3):362-370, 1970.

Renovascular hypertension in children may have a wide clinical spectrum, can sometimes present a difficult diagnostic problem, and should be excluded in any pediatric patient with otherwise unexplained hypertension. Of 3 children with renovascular hypertension, one, a 9½-year-old male, had neurofibromatosis and showed frank hypokalemia, polyuria, and hyponatremia. On examination the S was found to have bilateral renal artery stenosis. A second patient, a 16-year-old boy, had stenosis of 1 renal artery caused by fibromuscular hyperplasia. The third S, a 5½-year-old male, presented a complicated diagnostic problem and required repeated arteriograms and renal vein catheterizations for differential renin assay. An isolated malformation of one small intrarenal artery was observed. Cases such as the latter, in which metabolic and hormonal abnormalities persist in the absence of radiographic evidence of a lesion, point to the need for intense and selective study of the renal arterial system. (28 refs.) - *M. S. Fish.*

*San Francisco General Hospital
San Francisco, California 94110

MEDICAL ASPECTS – Etiologic Groupings

Prenatal influence

1914 **ARTO-MEDRANO, F.** Influence of cord around the neck on fetal acid-base balance in vigorous newborn infants. *American Journal of Obstetrics and Gynecology,* 107(7):1035-1043, 1970.

A comparison of acid-base balance in 14 fetuses born with umbilical cord around the neck and 14 controls shows the capillary and umbilical artery pH values to be lower in the group with cord around the neck. However, the good neonatal condition of these infants reflected a relatively normal umbilical vein pH. It is hypothesized that a moderate degree of compression impairs circulation through the neck vessels but does not affect umbilical circulation, and that the mild acidosis

may be explained on the basis of cephalic venous stasis. (12 refs.) - *E. L. Rowan.*

Maternidad Ciudad Sanitaria de la
Seguridad Social "Francisco Franco"
Barcelona, Spain

1915 **DEBDAS, ALOKE; & CHOWDHURY, N. N. ROY.** Fetal distress and pH value of umbilical cord blood. *American Journal of Obstetrics and Gynecology,* 107(7):1044-1046, 1970.

The average pH of both arterial and venous umbilical cord blood in 216 cases of fetal distress was lower than that of 40 normal controls. Fetal distress was defined as a heart rate greater than 160 or less than 100 beats per minute, grossly irregular cardiac rhythm, or passage of meconium. For distressed infants the average venous pH was 7.02, and the arterial pH 6.91, as compared with control values of venous pH 7.25 and arterial pH 7.19. More depressed infants were more acidotic, but it was noted that there appears to be a mild physiologic acidosis even in normal infants. (5 refs.) - *E. L. Rowan.*

R. G. Kar Medical College
Calcutta, India

1916 **BLOOMFIELD, DANIEL K.** Fetal deaths and malformations associated with the use of coumarin derivatives in pregnancy. *American Journal of Obstetrics and Gynecology,* 107(6):883-888, 1970.

A review of the literature associating fetal death and malformation with the use of oral anticoagulants (coumarin) during pregnancy reveals 16 case reports in 10 articles. In only 6 of these cases was the death hemorrhagic and thus attributable to coumarin toxicity. The therapeutic range was clearly exceeded in 4 of these. Two cases of nasal bridge hypoplasia are reported after coumarin use, but syphilis cannot be ruled out in one case. Because coumarin therapy is essential in patients with prosthetic heart valves, pregnancy can best be managed by not exceeding the therapeutic range of the drug and by switching from oral coumarin to heparin 1 month before term. Pregnancy is not contraindicated when oral anticoagulants are used wisely. (19 refs.) - *E. L. Rowan.*

Mt. Sinai Hospital
Cleveland, Ohio 44106

1917 **MOOSA, A.; & *DUBOWITZ, V.** Peripheral neuropathy in Cockayne's syndrome. *Archives of Disease in Childhood,* 45(243):674-677, 1970.

A recent study has reported the first case of peripheral nerve involvement in Cockayne's syndrome. The 20 previously reported cases of this syndrome, likely a recessively inherited disorder, were characterized by a variety of conditions including dwarfism, unusual facies, scaly erythematous dermatitis, optic atrophy and other visual disorders, joint contractures, intracranial calcification, osteoporosis, tremor, diminished lachrymation and sweating, renal lesions, undescended testes, and MR. The S was a full-term male child who, at 7 months, failed to continue to thrive although early developmental milestones were normal. On examination at 6 years 6 months he looked like a wizened old man and had many of the typical features of Cockayne's syndrome. Nerve conduction studies indicated that segmental demyelination due to Schwann cell damage was probably present, and sural nerve biopsy confirmed this diagnosis. These findings and other features which this syndrome has in common with metachromatic leukodystrophy – early normal development, followed by appearance of symptoms at 2-3 years – suggest that Cockayne's syndrome may be a leukodystrophy. (21 refs.) - *M. S. Fish.*

*The Children's Hospital
Sheffield, S10 2TH, England

1918 **LISTER, A. H.** Future for children with spina bifida. *Lancet,* 2(7680):982-983, 1970.

Although increasing attention is being given to the medical and surgical care of the growing number of children with spina bifida, large numbers of these children who are growing up still have physical defects. In his early years the child accepts his predicament and the care of parents. As the child matures, however, both he and his family experience new frustrations. These children face a particularly difficult life, and they should be given more assistance to develop the skills they

have. Social and educational progress can afford them some pleasure in being alive. (No refs.) - *B. J. Grylack.*

West Cumberland Hospital
Whitehaven, England

1919 MARTIN-BOUYER, G.; MONSET-COUCHARD, M.; AMIEL-TISON, C.; BOMSEL, F.; LARROCHE, J. -C.; DEHAN, M.; DOMMERGUES, J.-P.; ELCHARDUS, J.-F.; SAUDUBRAY, J.-M.; ZAMET, P.; & MINKOWSKI, A. La ventilation artificielle dans la maladie des membranes hyalines. Technique et resultats (Artificial ventilation in hyaline membrane disease. Technique and results). *Archives Francaises de Pediatrie*, 27(4):417-435, 1970.

Artificial ventilation was employed with 75 neonates diagnosed with hyaline membrane disease on the basis of clinical signs of respiratory distress and radiological indications. Sixty-seven were treated with the interval regulated respirator, 3 with the Bennet respirator, 3 with the Air-Shields incubator, and 2 with the incubator followed by the interval regulated respirator. Thirty-two infants were ventilated according to biological criteria, and 33 according to clinical criteria. In 10 cases, the 2 sets of criteria were present simultaneously. Of the 75 newborns, 30 survived. Comparison of survival rates appeared to favor the use of biological criteria. Among the surviving Ss, duration of ventilation in pure oxygen, duration of continuous ventilation, period of ventilation deprivation, and total duration of incubation were the 4 characteristics utilized to measure the success of ventilation. Through the adoption of pH equal to or greater than 7.30, arterial pressure of carbon dioxide ($PaCO_2$) equal to or less than 50 mm of mercury (Hg), and arterial pressure of oxygen (PaO_2) equal to or greater than 80 mm Hg as normalization criteria, it was possible to ascertain that the acid-base equilibrium was reestablished the most rapidly. This method is too recent to produce a significant neurologic evaluation at the present time. (24 refs.) - *B. J. Grylack.*

Centre de Recherches Biologiques Neonatales
75 - Paris 14^{e} France

1920 PONTE, C.; GAUDIER, B.; GUERRIN, F.; REMY, J.; BONTE, C.; LEQUIEN, P.; & FLINOIS, C. La ventilation assistee dans le traitement de la detresse respiratoire idiopathique et de l'inhalation amniotique du nouveau-ne (Assisted ventilation in the treatment of idiopathic respiratory distress and amniotic fluid inhalation in the newborn). *Archives Francaises de Pediatrie*, 27(1):73-84, 1970.

Sixty-four out of 371 newborns with idiopathic respiratory distress and 11 out of 14 newborns with massive amniotic fluid inhalation were treated by means of assisted ventilation. Indications and contraindications were posited on clinical bases. Of the 64 newborns with idiopathic respiratory distress, 31 died rapidly during the 62 hours following intubation, largely from progressive anoxia. Of the 13 newborns who died from other causes, 7 deaths were attributable directly or indirectly to the assisted ventilation technique. Twenty of the 64 infants survived. Six out of 11 newborns with amniotic fluid inhalation survived, 1 of whom manifested neurologic sequelae. Four out of the 5 who died had bilateral lesions. In general, 24 out of 75 newborns recovered without evident neurologic sequelae. It appears that assisted ventilation represents only the last therapeutic recourse after failure of hyperoxygenation and metabolic reequilibration. (19 refs.) - *B. J. Grylack.*

C.H.U. de Lille
Hopital Calmette
Lille, France

1921 ADAMS, FORREST H.; FUJIWARA, TETSURO; EMMANOUILIDES, GEORGE C.; & RAIHA, NIELS. Lung phospholipids of human fetuses and infants with and without hyaline membrane disease. *Journal of Pediatrics*, 77(5):833-841, 1970.

Total phospholipids, phosphatidylcholine (PC), and disaturated PC (surfactant) were measured and compared in lungs from 20 non-breathing abortuses, 14 infants with hyaline membranes, and 8 infants who died from causes other than hyaline membrane disease. Levels of total phospholipids and PC were stable during the first 6 months of gestation, while the amount of disaturated PC gradually increased. All 3 increased perceptibly between 6 and 9 months. Infants with hyaline membrane disease had values similar to those fetuses of 6 months' gestation. Surface tension

measurements expressed as stability index were also abnormally low in these infants. This is good evidence for a prenatal deficiency of surfactant in hyaline membrane disease. (21 refs.) - *E. L. Rowan.*

UCLA School of Medicine
Los Angeles, California

1922 **PRIMROSE, D. A.** Cervico-oculo-acoustic dysplasia. *British Medical Journal,* 3(5714):108, 1970.

A severely MR adult female shows evidence of cervico-oculo-acoustic dysplasia, a rare condition more commonly reported in children. The patient has Klippel-Feil syndrome, low hairline, small low-set ears, mutism, partial deafness, blindness, ocular paresis, optic atrophy, and epilepsy. (4 refs.) - *E. L. Rowan.*

Royal Scottish National Hospital
Larbert, Stirlings, Scotland

1923 **MILHORAT, THOMAS H.** Cerebrospinal fluid dynamics. *New England Journal of Medicine,* 283(14):763-764, 1970. (Letter)

A reply to a letter which gave alternatives to the explanations offered in a previous paper on cerebrospinal fluid dynamics states that since internal hydrocephalus was usually present in the patients studied, and obstructions were complete, the alternative explanation that surgical removal of the choroid plexus increased the obstruction or altered CSF flow from the ventricles to the arachnoid seems unlikely. (1 ref.) - *E. Kravitz.*

Children's Hospital
Washington, D. C.

1924 **CASTELLINO, RONALD A.; ZATZ, LESLIE M.; & DeNARDO, GERALD L.** Cerebrospinal-fluid dynamics. *New England Journal of Medicine,* 283(14):763, 1970. (Letter)

An explanation is offered for the observations of other workers on cerebrospinal fluid (CSF) dynamics in infantile obstructive hydrocephalus using ^{131}I-labelled serum albumin (RISA). RISA loss may have been caused by delayed ventricular passage and eventual absorption by the blood in the area of the sagittal sinus. The decreased ventricular disappearance rate of RISA after removal of the choroid plexus may have been due to additional blockage created by the surgery or to additional reduction of the subarachnoid space over the hemisphere, which would decrease CSF flow and absorption. (2 refs.) - *E. Kravitz.*

Stanford University Medical Center
Stanford, California

1925 **HOLMES, LEWIS B.** Norrie's disease—an X-linked syndrome of retinal malformation, mental retardation, and deafness. *New England Journal of Medicine,* 7(284):367-368, 1971.

Incorrect diagnoses of a family in which 6 males in 4 generations were affected with Norrie's disease reflect the general unawareness of this syndrome. Two difficulties in identifying the malady are the dramatic ocular changes with increasing age (the primary eye malformation is thought to be hypoplasia of the inner retinal layer) and the fact that not every victim has subnormal intelligence and defective hearing. While eye abnormalities are developing, subnormal psychomotor signs appear in 1/3 of patients. The disease has evidenced few neuropathological data, no consistent abnormalities, and no chromosomal or biochemical aberrations. Carrier females reveal no eye or hearing defects. Proper diagnosis will permit family counseling on the risks of having affected children. (5 refs.) - *B. Berman.*

Massachusetts General Hospital
Boston, Massachusetts

1926 **MacMAHON, BRIAN; & YEN, STELLA.** Unrecognised epidemic of anencephaly and spina bifida. *Lancet,* 1(7688):31-33, 1971.

Identification of children born with anencephaly and spina bifida as long ago as 1832 and 1835 has been made possible through records in delivery-books kept by the Boston Lying-In and the Providence Lying-In Hospitals. Rates of about 2 per 1,000 births, comparable to levels noted

since 1950, occurred during 1890-1920. A series of high rates in the 1920-1949 interval peaked during 1929-1932 at 3 times the rates before or after the epidemic. Failure to recognize the epidemic of neural-tube defects is attributed to its insidious onset and decline, and to clinical signs that aroused no new interest, since they were already familiar to obstetricians and pediatricians. It is of interest that the epidemic coincided with the Great Depression and the era of Prohibition, but various discrepancies obviate drawing any etiological conclusions. However, future etiological hypotheses should take this association into account, since it is nearly certain that environmental factors play a strong role in the development of these defects. (13 refs.) - *B. Berman.*

Harvard School of Public Health
Boston, Massachusetts

1927 Management of hydrocephalus. *Lancet,* 1(7699):587-588, 1971. (Editorial)

Analysis of the lactate and pyruvate content of cerebrospinal fluid (CSF) in hydrocephalic children has shown a metabolism decline, suggesting that tissue hypoxia from elevated intracranial pressure causes brain damage and MR. In multiple noncommunicating cranial cavities, it is desirable to have multiple shunts terminating in a single venous catheter. However, shunt treatment has many complications, such as bacteremia, in which case immediate shunt replacement is advisable. Embolic pulmonary complications can be detected chemically by radioactive lung scan. The absence of basic research in hydrocephalus is disappointing: it would seem quite possible to produce a drug to control development of hydrocephalus by controlling the CSF secretion rate. (No refs.) - *B. Berman.*

1928 Anencephaly. *Clinical Pediatrics,* 10(1):3-4, 1971.

The causes of anencephaly are unknown, but it has a close familial association with spina bifida; many cases of primary congenital hydrocephalus belong to the same etiologic grouping. Pediatricians must advise couples with an anencephalic infant that there is a tenfold risk of anencephaly or spina bifida in subsequent offspring; there is a great tendency for the same type of anomaly to recur. In one series of 498 index cases, there were 26 anencephalics in 887 viable siblings, 13 with spina bifida, and 5 hydrocephalics. Anencephaly is more common in girls. (9 refs.) - *B. Berman.*

1929 **SOLOWY, MARIAN K.; & SHEPARD, FRANK M.** Anencephaly in Central Virginia. *Clinical Pediatrics,* 10(1):43-45, 1971.

A 9-year retrospective review in Charlottesville, Virginia, showed 23 anencephalic infants among 23,381 deliveries; 21 were Caucasian, 2 Negro; 14 were girls. Mothers' ages ranged from 16 to 44 years (90% were less than 31 years); no mother had a history of anencephalic offspring. Four infants showed additional non-CNS congenital defects. Conception in the second and third quarters of the year showed some correlation with a higher incidence of anencephalic births. The sudden rise during 1969 suggested a new environmental factor, possibly the Hongkong influenza epidemic. There is need for careful obstetric records, especially during prenatal visits, with respect to illnesses and drugs taken during pregnancy. (20 refs.) - *B. Berman.*

University of Virginia School of Medicine
Charlottesville, Virginia

1930 **BUNDEY, SARAH; DUTTON, G.; & WELLS, R. S.** Tuberous sclerosis without adenoma sebaceum. *Journal of Mental Deficiency Research,* 14(Part 3):243-249, 1970.

A family which was observed was considered to be the first identified family in which a parent without adenoma sebaceum had 3 children with tuberous sclerosis. The father was 42 years old and had a shagreen patch on his left buttock. All 3 children were MR. It has been observed that shagreen patches occur only as part of the tuberous sclerosis complex. The absence of adenoma sebaceum does not exclude a diagnosis of tuberous sclerosis, since it may be a late manifestation. The described family illustrates a situation in which a shagreen patch has the same genetic significance as adenoma sebaceum. If the shagreen patch of the father had been discovered earlier, he might have received genetic counseling,

and the tragedy of bearing 3 MR children might have been averted. MR in heterozygotes for the gene tuberous sclerosis occurs in at least 2/3 of all cases. (13 refs.) - *B. J. Grylack.*

Institute of Child Health
London W.C. 1N 1EH, England

1931 **SHEAR, CAROL S.; NYHAN, WILLIAM L.; KIRMAN, BRIAN H.; & STERN, JAN.** Self-mutilative behavior as a feature of the de Lange syndrome. *Journal of Pediatrics,* 78(3):506-509, 1971.

Persistent self-mutilative biting which resulted in the loss of lip tissue was observed in 2 males who exhibited characteristic roentgenographic abnormalities and the dermatoglyphs of the de Lange syndrome. The first patient evidenced profound developmental retardation and had a Psyche-Cattell IQ of 13 at age 5-11 years. Self-mutilation involved biting and scratching various parts of his body and dislocating his hips. An intensified operative training program which included aversive stimuli was useful in management. In the second case, speech had not developed by age 5 years, and he had a developmental level between 12 and 18 months on the Piaget Sensorimotor Scale at that time. Self-mutilation involved picking his face, pulling his hair, and biting his lower lip. Management involved the removal of the 12 frontmost teeth. Self-mutilative behavior may be a distinctive feature of the de Lange syndrome. (8 refs.) - *J. K. Wyatt.*

University of California
La Jolla, California 92037

1932 **STRATFORD, BERNICE F.** Abnormalities of early human development. *American Journal of Obstetrics and Gynecology,* 107(8):1223-1232, 1970.

Of 254 human conception products studied, 159 were abnormal (28 of 44 extrauterine, 131 of 210 intrauterine); 63 of 158 embryos showed maldevelopment. Most specimens were less than 9 weeks old. Abnormal conceptuses were of 2 main types: In one, there was virtually no embryo; there was defective placental tissue and gross pathology throughout. In the second, there were embryos with specific localized abnormalities. Of 131 intrauterine abnormalities, 57 were local and 74 were gross. Of 28 extrauterine abnormalities, 6 were local and 22 gross. (39 refs.) - *E. Kravitz.*

University of Melbourne
Melbourne, Victoria, Australia

1933 **SACREZ, R.; GIGONNET, J.-M.; STOLL, C.; GROSSHANS, E.; & STOEBNER, P.** Encephalopathie congenitale et dyschromie familiale (Congenital encephalopathy and familial dyschromia). *Archives Francaises de Pediatrie,* 27(9):995, 1970.

Four observations were made of the incontinentia pigmenti achromians syndrome. Three sisters aged 4 years, 28 months, and 17 months presented with a congenital encephalopathy with profound MR, marked axial hypotonia, quadriparesis with pyramidal signs, craniofacial dysmorphia and osteodental anomalies, partial anodontia, irregularity and quadrangular projection of the center of the dorsolumbar vertebral plateaus, and achromic spots on the abdomen and lumbar region which progressed to the upper section of the trunk and the base of the limbs. The mother, who showed slight mental debility, had dysmorphic scars and a discrete achromic dermatosis similar to that of her daughters. This syndrome and Bloch-Sulzberger disease are both characterized by exclusive appearance among females, familial occurrence, and similar neuroectodermic dysplasias. The pigmentary aspect of the Bloch-Sulzberger disease and the achromic dermatosis of the incontinentia pigmenti syndrome are probably 2 successive manifestations of the same pathological process. (No refs.) - *B. J. Grylack.*

No address

1934 **DORMOY, O.; & AJZENBERG, D.** Encephalopathie familiale avec oligophrenie et indifference congenitale a la douleur et syndrome de Laurence-Moon-Bardet-Biedl (Familial encephalopathy with oligophrenia and congenital indifference to pain and the Laurence-Moon-Bardet-Biedl syndrome). *Annales Medico-Psychologiques,* 2(4):545-564, 1970.

An 8-year-old girl presented signs which were consistent with the Lesch-Nyhan syndrome.

Investigation of the level of hypoxanthine-guanine-phosphoribosyl-transferase, however, could not confirm the diagnosis of this syndrome. The presence of an encephalopathy and the absence of thermal sensibility favored the diagnosis of congenital indifference to pain. A sister of the S manifested the Laurence-Biedl syndrome, characterized primarily by polydactylism, progressive evolution of blindness and obesity, MR, radiologically diagnosed anomalies, and a morphologically abnormal urinary tract. In view of the existence of 2 rare and apparently disparate diseases within the same family, an association was sought. Upon examination, alkaline phosphatase levels were found to be elevated in both sisters, although no numerical or structural abnormalities of the chromosomes were identified. Another pathological finding was an increase in esterases in both the S and her father. The father also presented with an increase in lymphocytic polysaccharides, probably connected with an anomaly of glucosidic metabolism. While definite conclusions could not be drawn from the existence of various pathological findings in the family, the fact that the absence of morphological chromosomal anomalies does not preclude their functional deterioration should be emphasized. (No refs.) - *B. J. Grylack.*

Clinique de Psychiatrie du C.H.R.
Universite Paul-Sabatier
Toulouse, France

1935 **STERNBERG, B.; MISES, J.; & LERIQUE-KOECHLIN, A.** E.E.G. et hydrocephalies neo-natales. Bilan de 50 cas (EEG and neonatal hydrocephalus. Report of 50 cases). *Revue Neurologique,* 122(6):491-493, 1970.

An investigation was made of 2 groups of 25 children with an early diagnosis of hydrocephalus whose initial EEG had been recorded before the age of 3 months. Ss were divided into 4 categories according to the characteristics of their first EEG. In the case of Ss manifesting a normal or overly active baseline without distinct anomalies, an EEG with localized anomalies, or a globally altered EEG with or without crises, a valid correlation was found with a distinct etiology, a history of prematurity, the presence of crises, or the extent of psychomotor development. Ss manifesting an asymmetrical EEG with hemispheric or localized inactivity or hypoactivity did so in association with an agenesis of the corpus callosum in 1 case, with a hemispheric tumor in 1 case, and with subdural hematomas in 3 cases. Thus, a definite relationship was noted between EEG inactivity and asymmetry, on the one hand, and the probability of a tumor, malformation, or subdural hematoma, on the other. Normal tracings during evolution of the EEG were shown to be consistent with the possibility of normal development, a finding which may ascribe a prognostic value to the EEG. (No refs.) - *B. J. Grylack.*

L'Hopital Saint-Vincent-de-Paul
F. 75-Paris 14^{e} France

1936 **VERGER, P.; LEMAN, P.; COHADON, F.; GUILLARD, J.-M.; MALLARIVE, J.; & SAINT-JAMMES, TORTES.** Le devenir des nourrissons hydrocephales operes par derivation ventriculo-atriale. Une nouvelle serie de 32 cas (The course of hydrocephalic infants treated with ventriculo-atrial shunt. A new series of 32 cases). *Archives Francaises de Pediatrie,* 27(8):859-860, 1970.

Thirty-two hydrocephalic infants observed between 1964 and 1968 were treated with a ventriculoatrial shunt with the Pudenz valve. Seventy percent of the infants had no vomiting but manifested serious neurological signs in addition to an increase in head circumference. In the immediate postoperative period, 2 cases of transitory intracranial hypotension were revealed. Twenty-five percent showed fluid leakage, and 25%, including the same Ss in some cases, revealed obstructive crises. Sixteen infants were treated only once, while 16 were retreated as a consequence of the noted complications. The results obtained from the 25 surviving infants showed that craniofacial morphology was normal in 50%. Sixty percent manifested no appreciable aftereffects of hydrocephaly, but half of these Ss had no evident preoperative neurological deficit. The results of testing by the Gesell Test up to age 3 and the Termann-Merill Test, form L, from then on were found to be ambiguous. (No refs.) - *B. J. Grylack.*

No address

1937 **BATTIN, J.; & MARCHAND, J.-CL.** Syndrome de Prader, Labhardt et Willi

(The Prader, Labhardt, and Willi syndrome). *Archives Francaises de Pediatrie*, 27(8):861-862, 1970.

The Prader-Labhardt-Willi syndrome has been reported in a male, aged 4½ years. The S was born without particular difficulties but remained very hypotonic during the postnatal period. Psychomotor retardation was signified by inability to raise the head until 4 months, inability to sit until 1 year, and inability to walk until 22 months. The S had language, although it was agrammatical, and he was able to attend kindergarten. Bulimia, obesity, and genu valgum appeared at approximately age 3. The child had a small penis, bilateral cryptorchidism, scrotal hypoplasia with a hollow median raphe that appeared to be pseudovalvular, extensive cavities, and strabismus. Endocrine function, glucose metabolism, and chromosomal analysis were normal. (No refs.) - *B. J. Grylack.*

No address

1938 **BATTIN, J.; AZANZA, X.; & ALBERTY, J.** Syndrome de Prader-Willi-Labhardt chez une fille avec evolution d'un diabete insulinoprive (The Prader-Willi-Labhardt syndrome in a girl with a course of insulinopenic diabetes). *Archives Francaises de Pediatrie*, 27(8):862-863, 1970.

The appearance of diabetes mellitus as a late and inconstant feature constitutes a new element in the description of the Prader-Willi-Labhardt syndrome. A girl hospitalized at age 7 with encephalopathy was found to have glycosuria which was part of a pattern similar to that of insulinopenic diabetes in infants. The maternal grandfather of the S had also been obese and diabetic. Hydramnios during pregnancy and birth at term at a weight of 3,200 g were followed by postnatal respiratory difficulties as well as apathy and hypotonia during the first 6 months of life. Bulimia and obesity were evident from the first year onward. At age 7, the S weighed 33 kg and had a height of 1.30 m, corresponding to a weight age of 11 years. Bone age was normal. The S had a dysmorphic and inexpressive face, numerous cavities, genu valgum, and strabismus. The existence of non-ketoacidotic diabetes with obesity favored treatment by a restrictive diet along with tolbutamide administration, which resulted in a loss of 8 kg in 7 months and increasing frequency of acetonuria. In the 3 months following a second course of tolbutamide, the S lost an additional 2 kg, edema from hypoproteinemia was apparent, and acetonuria became permanent. Tolbutamide was stopped, and insulin administration was initiated to counter this intractable evolution of the Prader syndrome. (No refs.) - *B. J. Grylack.*

No address

1939 **BODA, D.; MURANYI, L.; ALTORJAY, I.; & VERESS, I.** Peritoneal dialysis in the treatment of hyaline membrane disease of newborn premature infants. *Acta Paediatrica Scandinavica*, 60(1):90-92, 1971.

Peritoneal dialysis was performed on 50 premature infants with hyaline membrane disease to correct metabolic disturbances without excessive quantities of fluid and electrolytes and to eliminate toxic metabolic products that may accumulate during hypoxia. A control group of 42 infants received conventional treatment of alkali glucose infusion. No respiratory distress or other serious complications were noted in the infants treated with dialysis, and 21 of the 50 infants recovered, as compared with 7 of the 42 treated with alkali and glucose. Further clinical experiments will use arterial oxygen tension measurements to investigate the therapeutic effects of peritoneal dialysis. (6 refs.) - *N. Jarvis.*

Department of Pediatrics
University of Szeged, Hungary

1940 **DRILLIEN, C. M.** Prognosis of infants of very low birth-weight. *Lancet*, 1(7701):697, 1971. (Letter)

Long-term followup of very low birth weight infants has suggested differential prognosis based on the etiology of the condition. Infants who are delivered prematurely and/or are of low birth weight because of developmental abnormalities show a high incidence of congenital anomalies and severe MR. Small-for-dates infants malnourished or hypoxic during the third trimester show only mild MR and neurological impairment. Premature infants of appropriate weight are potentially normal with modern methods of premature-baby care. (3 refs.) - *E. L. Rowan.*

University of Edinburgh
Edinburgh, EH9 IUW, Scotland

1941 Epidemics of anencephaly and spina bifida. *Lancet*, 1(7701):689-690, 1971. (Editorial)

Neural tube defects (anencephaly and spina bifida) appear to occur in epidemics. Changes in diagnostic selection or population composition do not account for the peak incidence of these anomalies in Boston, Massachusetts, and Providence, Rhode Island from 1932-1934; England and Wales in 1942-1943, and 1954-1955; and Atlanta, Georgia, Vermont, and Philadelphia, Pennsylvania, in 1962. Identification of significant environmental factors might lead to prevention. (11 refs.) - *E. L. Rowan.*

1942 NAGGAN, LECHAIM. Anencephaly and spina bifida in Israel. *Pediatrics*, 47(3):577-586, 1971.

Neural tube defects (anencephaly and spina bifida) were noted in 776 of the 531,985 infants born to Jewish mothers in Israel between 1958 and 1968. An unusually high percentage (44.1%) of the anencephalic infants were male. Rates for both anomalies were markedly higher after the fifth child. Immigrants from Iran, Iraq, and Yemen had the highest risk. Rates of spina bifida decreased during the study period. There was no seasonal variability. The downward trend is suggestive of environmental causation. The decrease in spina bifida may be due to improving socioeconomic status, but the effects of birth order and ethnic origin in anencephaly change more slowly. (23 refs.) - *E. L. Rowan.*

17 Emek Dotan TelHashomer
Ramat Gan, Israel

1943 NEU, RICHARD L.; KAJII, TADASHI; GARDNER, LYTT I.; NAGYFY, STEPHEN F.; & *KING, SADDIE. A lethal syndrome of microcephaly with multiple congenital anomalies in three siblings. *Pediatrics*, 47(3):610-612, 1971.

Three siblings exhibited a syndrome of microcephaly and multiple congenital anomalies of the limbs, described as flexion deformities. The first pregnancy had ended in abortion during the first trimester. Karyotypes of one of the infants and parents were normal. An autosomal recessive mode of inheritance is postulated. (1 ref.) - *E. L. Rowan.*

*S.U.N.Y. Upstate Medical Center
Syracuse, New York 13210

1944 OKUBOVEJO, A. Extra leg with anencephaly. *Lancet*, 1(7691):188, 1971. (Letter)

A three-legged male child was born at the State Hospital, Ijebe-Ode, Nigeria. The infant was alive but blue at birth and weighed 8 lbs. 2 oz. He had 2 normal legs 8 inches long and a third arising from the back just above the cleft of the buttocks. The third leg was 2 inches shorter than the others and had only 2 toes. The face was normally developed as far as the hair-line, but there the skull stopped abruptly and the poorly developed brain, covered by meninges, protruded in a cauliflower manner. The mother, a primapara aged 17 and a native of the area, had used during the first trimester a folk medicine commonly taken by pregnant women in that part of the country. She had not been ill during that period. At the Rural Health Center she had been given the usual antenatal drugs—ferrous sulphate, vitamin C, and folic acid. (No refs.) - *J. C. Moody.*

State Hospital
Ijebu-Ode, Nigeria

1945 ELWOOD, J. H.; & MACKENZIE, G. Comparisons of secular and seasonal variations in the incidence of anencephalus in Belfast and four Scottish cities, 1956-66. *British Journal of Preventive & Social Medicine*, 25(1):17-25, 1971.

The secular and seasonal variations in the incidence of anencephalus occurring in all live births and stillbirths of 28 or more weeks of gestation in Belfast, Aberdeen, Dundee, Edinburgh, and Glasgow were examined over the period 1956-1966. The incidence showed a small overall increase in Belfast, no change in Glasgow, and a decrease in Aberdeen, Dundee, and Edinburgh. A winter excess of anencephalic births occurred in all cities except Aberdeen, where there was an excess in summer. Using a multivariate regression model, the associations between the monthly

number of anencephalic births and 4 monthly climatic indices (maximal and minimal air temperatures, hours of sunshine, and total rainfall) were determined with data for Belfast, Edinburgh, and Glasgow. Low but significant negative correlations with maximal air temperature occurring during the first month of gestation were found in Belfast and Glasgow but not in Edinburgh. It is concluded that variations in climate are associated with only a small proportion of the seasonal variations in the incidence of anencephalus and that a major direct deleterious influence of climatic factors on the central nervous system of the 1-month old fetus cannot be substantiated. (33 refs.) - *J. C. Moody.*

Queen's University
Belfast, Northern Ireland

1946 **HERBST, BERNADETTE A.; & COHEN, MICHAEL E.** Linea nevus sebaceus. *Archives of Neurology*, 24(4):317-322, 1971.

A fourth case of a new neurocutaneous syndrome occurred in a 5½ month old child who presented with skin lesions in a characteristic midline distribution. Histological findings were consistent with the diagnosis of nevus sebaceus of Jadassohn. There was also sudden and almost simultaneous onset of psychomotor deterioration with onset of seizures 3 weeks prior to admission. The electroencephalogram revealed a typical hypsarrythmic pattern, a feature not previously reported. Since both the skin and the nervous system are derived from the primordial ectoderm, the time of skin lesion may indicate when the time of insult took place. The frequent association of abnormalities of the nose with congenital defects of the brain indicates that the nevi along the midline of the nose may be the visible manifestation of underlying cerebral disturbances and thus pinpoint in which developmental period this malformation took place. (20 refs.) - *S. Glinsky.*

Children's Hospital of Buffalo
Buffalo, New York

1947 **RAWLINGS, GRACE; REYNOLDS, E.O.R.; STEWART, ANN; & STRANG, L. B.** Changing prognosis for infants of very low birth weight. *Lancet*, 1(7698):516-519, 1971.

Due to much improved medical and nursing care infants of very low birth weight now seem to have a greatly improved prognosis for survival and normality. The Ss were 68 infants of less than 1500g birth weight, born during the years 1966 to 1969, whose progress has been followed up. Hospital care emphasized prevention of biochemical and other abnormalities, while home care involved frequent clinical examinations and developmental screening. At a mean conceptual age of 2 years 3 months, with a range of 9 months to 4 years 3 months, 59 (86%) appear to be normal, 5 (7.4%) are abnormal, and 4 (5.9%) are classified as doubtful. The application of modern methods of care has substantially bettered the prognosis of such infants and appears to invalidate earlier findings in this area. (23 refs.) - *S. Glinsky.*

University College
London, England

1948 Development and very low birth weight. *Lancet*, 1(7698):534-535, 1971.

Recent success in increasing the survival rate of infants of very low birth weight and reducing abnormality warrants more intensive study of these phenomena and a reversal of earlier pessimistic attitudes. Until very recently, studies predicted an increase of abnormality as medical knowledge increased the survival rate of very low birth weight infants. A study in a London hospital has reversed such predictions by means of an intensive-care program aimed particularly at preventing insults to the brain, such as hypoxia, low blood sugar, and raised bilirubin levels. Although these results are not fully conclusive, they should lead to a change in attitude and policies regarding such follow-up studies. Considering the maternal deprivation occasioned by such studies, they should also assess this factor's effect as well as that of perinatal chemical disturbances on intellectual impairment. (4 refs.) - *S. Glinsky.*

1949 **DAMYANOV, I.; & DUTZ, W.** Anencephaly in Shiraz, Iran. *Lancet*, 1(7689):82, 1971. (Letter)

Although there is evidence that environmental factors play a pathogenic role in anencephaly, ethnic background seems to be more important. The incidence of anencephaly was analyzed from

the necropsy files from the Pahlavi School of Medicine in Shiraz, Iran. These files cover approximately half the children born in Shiraz hospitals. Out of 14,520 deliveries during the period from August, 1966 to August, 1970, 23 anencephalic children were born. Most of the children were born in December and 13 were girls. The incidence of 0.16% is comparable with the 0.121% reported for non-Ashkenazy Israelis and suggests an ethnic explanation. (5 refs.) - *S. Glinsky.*

Pahlavi University School of Medicine
Shiraz, Iran

1950 ROBINOW, MEINHARD; PFEIFFER, RUDOLF A.; GORLIN, ROBERT J.; McKUSICK, VICTOR A.; RENUART, ADHEMAR W.; JOHNSON, G. FRANK; & SUMMITT, ROBERT L. Acrodysostosis. *American Journal of Diseases of Children*, 121(3):195-203, 1971.

Acrodysostosis, a new congenital malformation syndrome, is described on the basis of 9 new cases and 11 cases from the literature. The major features of this rare syndrome are peripheral dysostosis, nasal hypoplasia, mental deficiency, and growth failure. Additional manifestations include premature skeletal maturation in hands, feet, and elbows and some less constant abnormalities of the spine, mandible, and skull. Hypogenitalism, middle ear infections, and an abundance of pigmented nevi may be manifested occasionally. The condition is usually recognized at birth or during the first year, and deformities are slowly progressive during the growth period. Neither familial incidence nor parental consanguinity has been encountered, and no biochemical defect has been demonstrated. (22 refs.) - *S. Glinsky.*

Children's Medical Center
Dayton, Ohio

1951 ROHLFING, BARBARA; LEWIS, KAY; & SINGLETON, EDWARD B. Rubinstein-Taybi syndrome. *American Journal of Diseases of Children*, 121(1):71-74, 1971.

An unusual case of Rubinstein-Taybi syndrome with a rare keloid formation is described. The patient is a 26-year-old MR white man and presents with cleft palate, double rows of teeth, associated deformities of the knees, elbow, and toenail, extensive keloid formation, aminoaciduria, and a history of maternal first trimester rubella. The findings of patellar and radial head dislocation may be those of chance association, as may be the historical rubella infection. However, the latter should be sought as a possible etiology of future Rubinstein-Taybi syndrome cases. The significance of the aminoaciduria is uncertain at present. Keloid formation has been found in only 2 other cases and is a generally unrecognized minor characteristic of the syndrome. (5 refs.) - *S. Glinsky.*

Texas Children's Hospital
Houston, Texas 77025

1952 WINICK, MYRON. Cellular changes during placental and fetal growth. *American Journal of Obstetrics and Gynecology*, 109(1):166-176, 1971.

Growth may be defined simply as an increase in the size of an animal or individual organ. In the first phase of growth, there is a proportional increase in weight, protein, and deoxyribonucleic acid (DNA) content (hyperplasia). The second phase is characterized by an increase in DNA that is slower than the increase in protein and weight (hyperplasia and concomitant hypertrophy). In the third stage, no further increase in DNA content takes place, and net protein and weight continue to increase at the same rate (hypertrophy). Although the placenta is largely a syncytium, its pattern of growth resembles that of organs made up of cells with discrete boundaries. Evidence in animals demonstrates that severe maternal protein restriction will curtail cell division in all fetal organs, including the brain. The nature of a stimulus which interferes with growth appears to be less important than the time during which the stimulus is active. Limited data suggest that children who weigh less than 2,000g at birth will sustain greater cellular damage than larger infants when both are subsequently exposed to postnatal malnutrition. The extent of nutritional deprivation necessary to produce certain changes is unknown at the present time. (30 refs.) - *B. J. Grylack.*

Cornell University Medical College
New York, New York

1953 **DANCIS, JOSEPH; & SMITH, ALFRED A.** Familial dysautonomia. *Journal of Pediatrics,* 77(1):174-175, 1970.

A recommendation that children previously classified as cases of congenital sensory neuropathy be included in the diagnostic category of familial dysautonomia, type II, does not appear to be justified on the basis of the description of the particular cases involved. While resembling those with familial dysautonomia in their sensory defect, these cases presented no evidence of parasympathetic insufficiency; secretion of tears was normal; and methacholine did not induce miosis or alter taste thresholds. Normal fungiform papillae and taste buds were also present. The term familial dysautonomia as now employed indicates a sharply defined group of children in terms of objective criteria, clinical history, and genetics. In the present state of knowledge, congenital sensory neuropathy likely should be retained as the more appropriate term for the cases described. (3 refs.) - *M. S. Fish.*

No address

1954 **HOLDEN, KENTON R.; & ALEXANDER, FRED.** Diffuse neonatal hemangiomatosis. *Pediatrics,* 46(3):411-421, 1970.

Steroid therapy proved to be ineffective in altering the course of diffuse neonatal hemangiomatosis in 2 infants. One infant, premature, cyanotic, with mild to moderate respiratory distress at birth and with blood in the first urine specimen, died at 18 hr despite administration of antibiotics, fresh whole blood, and cortisone. The observed thrombocytopenia was considered to result from platelet utilization in thrombosis of the hemangiomas in the heart, lungs, liver, spleen, kidneys, and cerebellum. Histological examination disclosed these to be thin-walled, with no obvious proliferation of endothelial cells or arteriovenous communications. The second case had more widely diffuse lesions and, after the onset of neurological deterioration at 2 weeks, succumbed at 35 days to extensive CNS involvement including hydrocephalus from aqueductal compression by hemangiomas. Involvement also included the skin, mucous membranes, gastrointestinal tract, lungs and pleura, thymus, mesentery, bladder, lymph nodes, larynx, heart, skeletal muscle, uterine cervix, iris, spinal cord, and meninges. The vascular lesion in this disease is possibly secondary to congenital absence of vessel wall pericytes. (17 refs.) - *M. S. Fish.*

NINDS, Clinical Center, NIH,
Bethesda, Maryland 20014

1955 **NITZAN, MENACHEM; & GROFFMAN, HELEN.** Hepatic gluconeogenesis and lipogenesis in experimental intrauterine growth retardation in the rat. *American Journal of Obstetrics and Gynecology,* 109(4):623-627, 1971.

Delay in the postnatal development of hepatic gluconeogenesis may contribute to susceptibility to neonatal hypoglycemia. Intrauterine growth retardation (IGR) in 23 rats was induced by ligating the artery supplying one uterine horn on the 17th day of gestation. The artery of the opposite horn was left untouched, permitting normal development of control fetuses on this side. *In vitro* studies with liver slices 3 hours after spontaneous delivery showed impaired glucose production from labeled alanine in the dysmature newborn rats. IGR may therefore be associated with a functional delay in the postnatal development of hepatic gluconeogenesis. Hepatic phospholipogenesis was also less active in the IGR fetuses. This may reflect developmental immaturity of membranous systems, which play an important role in the control and integration of hepatic metabolism. Reduction of placental blood flow might be responsible for IGR in humans as well. (25 refs.) - *S. Glinsky.*

Northwestern University Medical School
Chicago, Illinois 60611

1956 **LUGO, GUSTAVO; & CASSADY, GEORGE.** Intrauterine growth retardation. *American Journal of Obstetrics and Gynecology,* 109(4):615-622, 1971.

The intrauterine growth retarded (IGR) population constitutes a particularly vulnerable group in which quantitative and qualitative differences in morbidity and mortality are apparent. The Ss were 233 IGR infants delivered during a one-year period whose clinicopathological perinatal characteristics were contrasted with those of 2,814 normally grown neonates delivered at the same

institution during the same time interval. Perinatal mortality in the IGR group was 94.4 per 1000, while that for normal neonates was zero. In the surviving IGR infants the perinatal morbidity disturbances included significant incidences of perinatal asphyxia, congenital malformation, hypoglucosemia, and polycythemia. Although 73% of the IGR group was Negro, no racial predilection for IGR was demonstrated, but IGR may well be related to socioeconomic, nutritional or environmental conditions. The predictable spectrum of problems and the extraordinary risk of perinatal death in IGR infants point up the need for obstetricians to appreciate these risks and the vulnerability of this group. (18 refs.) - *S. Glinsky.*

University of Alabama Medical Center
Perinatal Research Laboratory
Birmingham, Alabama

1957 **FORT, ARTHUR T.** Adequate prenatal nutrition. *Obstetrics and Gynecology,* 37(2):286-288, 1971. (Editorial)

The nutritional supply line to the fetus should be kept full to avoid the consequence of an irreversible developmental handicap. The single, best indicator of reproductive success in terms of survival is birthweight. Long-term intellectual performance and general metabolic efficiency are also affected by birthweight. Low birthweight is associated with deficits in all these areas. (13 refs.) - *J. K. Wyatt.*

University of Tennessee College of Medicine
Memphis, Tennessee 38103

1958 Hydrocephalus with dementia. *New England Journal of Medicine,* 283(12):654-655, 1970. (Editorial)

The therapeutic value of ventriculovenous shunting in certain cases of dementia, e.g., hydrocephalus with normal CSF pressure, is discussed; diagnostic aids, e.g., pneumoencephalography and radioiodinated serum albumin (RISA) scanning, are summarized. RISA presents early ventricular concentration of radioactivity and its persistence and a lack of radioactivity over the cerebral convexities. Pneumoencephalographic findings include enlarged anterior lateral ventricles, narrowed corpus callosal angle, and lack of air over the hemispheres. (3 refs.) - *E. Kravitz.*

1959 **ZACHARY, R. B.** Coordinated care of the child with spina bifida. *Lancet,* 2(7677):824, 1970. (Letter)

The importance of an adequately conducted combined follow-up clinic in the management of children with spina bifida is stressed. Each hospital or group of hospitals dealing with this condition should arrange for the child and parent to consult all specialists during a single visit. (1 ref.) - *E. Kravitz.*

Children's Hospital
Sheffield S.10 2TH, England

1960 **LEBENTHAL, EMANUEL; SHOCHET, SOLLY BARNARD; ADAM, AVINOAM; SEELENFREUND, MOSHE; FRIED, AMNON; NAJENSON, THEODORE; SANDBANK, URIEL; & MATOTH, YEHUDA.** Arthrogryposis multiplex congenita: Twenty-three cases in an Arab kindred. *Pediatrics,* 46(6):891-899, 1970.

Twenty-three individuals belonging to 1 Arab kindred who all had clinical manifestations of arthrogryposis multiplex congenita (AMC) were studied. Clinically, the condition was present at birth, and the prominent features were contractures, primarily of the flexion type. The muscles at the affected joints were found to be weaker than those at normal joints, and the contractures of the big joints were of the mixed type, the flexion deformity being the most disabling. Six patients had congenital heart disease, and 1 was SMR. Electromyographic investigation of the Ss failed to reveal any pathological findings of neurogenic origin. Histologic and biochemical findings suggested a myogenic origin of the disease. The high concentration of persons afflicted with this rare congenital disease in a single kindred indicates that the condition is inherited myogenically and that it is probably due to homozygosity for an autosomal recessive gene. This form of congenital contractures can be contrasted with a neurogenic AMC which is determined by a dominant gene. (48 refs.) - *B. J. Grylack.*

Stanford University Medical Center
Stanford, California 94305

1961 **PASHAYAN, H.; LEVY, E. P.; & FRASER, F. C.** Can the de Lange syndrome always be diagnosed at birth? *Pediatrics*, 46(6):940-942, 1970.

In contrast to most cases of the de Lange syndrome, who look alike at birth, a patient was observed who had features suggestive of Turner's syndrome at the age of 9 months and the typical appearance of the de Lange syndrome at the age of 21 months. Stigmata of the de Lange syndrome include physical and mental retardation, synophrys, long curved eyelashes, low forehead, wide and depressed nose bridge, anteverted tip of nose exposing the nostrils, long filtrum, and small receding chin. Other anomalies exist in the syndrome but with lesser frequency. The late development of the characteristic features of the syndrome in this patient indicates that the accumulation of features characteristic of this and other syndromes varies not only from patient to patient but also within the same patient at different times. (7 refs.) - *B. J. Grylack.*

2300 Tupper Street
Montreal 108, P.Q., Canada

1962 **BERGNER, LAWRENCE; & SUSSER, MERVYN W.** Low birth weight and prenatal nutrition: An interpretative review. *Pediatrics*, 46(6):946-966, 1970.

Low birth weight is well established as an antecedent of excess mortality in infants and of MR and other neurological and psychological disorders. The lower the birth weight, the greater is the perinatal death rate and the higher is the incidence of brain disorders and MR. In New York City, the distribution of birth weight could account for the entire excess of perinatal deaths among black babies as compared with white babies. Data suggest that the uterine environment has an effect on fetal growth and that the measurable effects occur in late pregnancy. The effect of nutritional deficiency during pregnancy on the weight of offspring has been demonstrated in animal experiments, although data on humans are more equivocal. A fresh experiment aimed at preventing low birth weight by means of dietary supplements seems to be the most practical means of penetrating the web of associations between socioeconomic conditions and low birth weight. The ultimate concern of supplementation should not be the low birth weight itself but its postulated consequences for perinatal deaths, damage to the nervous system, and mental function and development. (81 refs.) - *B. J. Grylack.*

Department of Health
New York, New York

1963 **QUEENAN, JOHN T.; & GADOW, ENRIQUE C.** Polyhydramnios: Chronic versus acute. *American Journal of Obstetrics and Gynecology*, 108(3):349-355, 1970.

Of the 86,301 consecutive deliveries at the New York Hospital-Cornell Medical Center during the 20-year period from 1948 to 1967, there were 358 cases of polyhydramnios for an incidence of 0.41%. Among these cases, 122 (34%) were idiopathic, 88 (24.6%) were associated with maternal diabetes, 71 (19.8%) with congenital malformations, 41 (11.5%) with erythroblastosis fetalis, and 30 (8.4%) with multiple gestation. Only 6 patients (1.7%) fulfilled the strict criteria for acute polyhydramnios, which is a rampant fulminating process terminating in spontaneous labor, usually before the end of the second trimester. Aggressive management of chronic polyhydramnios by amniocentesis is indicated only for extreme maternal symptoms, whereas multiple amniocenteses should be used more readily in acute polyhydramnios if it is idiopathic or due to multiple gestation. (7 refs.) - *B. J. Grylack.*

New York Hospital–Cornell Medical Center
New York, New York

1964 **HOWAT, J. M.** Urinary ascites complicating spina bifida. *Archives of Disease in Childhood*, 46(245):103-105, 1971.

A female infant born with a lumbosacral meningomyelocele, hydrocephalus, and urinary ascites was successfully treated. The meningomyelocele was closed by surgery. The child had a normal upper urinary tract, but a cystogram disclosed a leak of contrast medium from the fundus of the bladder into the peritoneal cavity. Although the site of the leak could not be located, insertion of a suprapubic catheter resulted in satisfactory drainage. A holter valve was used to establish ventriculolatrial drainage on the fourth day of life. At the age of 6 months the child showed no

renal impairment. The absence of back pressure effects on the upper urinary tract suggests that the leak probably occurred a considerable time prior to birth. (7 refs.) - *J. K. Wyatt.*

Hospital for Sick Children
London WC1, England

1965 **PICKERING, DOUGLAS; LASKI, BERNARD; MacMILLAN, D. C.; & ROSE, VERA.** "Little leopard" syndrome. *Archives of Disease in Childhood,* 46(245):85-90, 1971.

The meaning of the 'r' in 'leopard' used to describe the 'little leopard' syndrome should be altered to represent MR. A report on 3 cases and a review of 24 others indicates that a significant number (7) were mentally as well as physically retarded. The 'little leopard' syndrome is a genetic condition inherited as an autosomal dominant which affects several systems, especially those derived from the ectoderm and mesoderm. Its features include short stature, lentignes, electrocardiograph and ocular defects, pulmonary infundibular stenosis, abnormal genitalia, MR, and deafness. (11 refs.) - *J. K. Wyatt.*

Radcliffe Infirmary
Oxford, England

1966 **CROALL, J.; & GRECH, P.** Radiological maturity of the small-for-dates fetus. *Journal of Obstetrics and Gynaecology of the British Commonwealth,* 77(9):802-807, 1970.

Singleton pregnancies with small-for-dates fetuses which were X-rayed *in utero* were selected from the records of 11,000 consecutive deliveries in a hospital maternity unit and were reviewed in order to determine the relationship between ossification delay and fetal growth retardation. In infants with birthweights more than 100g above the fifth percentile but within the twenty-fifth percentile, no evidence of ossification delay was revealed. In infants with birthweights from 100g above to 150g below the fifth percentile, ossification delay was suggested by the reduced number of lower femoral centers identified at 36 weeks onwards. In infants with birthweights 200g or more below the fifth percentile, delay in the appearance and growth of ossification centers was marked and resulted in misleading underestimates of maturity in half of the cases. In these grossly small-for-dates fetuses, reliable radiological verification of maturity can be obtained only when a lower femoral epiphyseal center is seen at 36 to 38 weeks. (13 refs.) - *B. J. Grylack.*

Northern General Hospital
Sheffield, England

1967 **CROALL, J.; & GRECH, P.** Hyperflexion of the small-for-dates fetus. *Journal of Obstetrics and Gynaecology of the British Commonwealth,* 77(9):808, 1970.

The incidence and significance of hyperflexion and the radiological maturity were assessed in 93 fetuses of low birthweight for dates. No hyperflexion was shown in 21 fetuses whose corrected birthweights were more than 100g above the fifth percentile. In 25 fetuses whose corrected birthweights ranged from 100g above to 150g below the fifth percentile, 2 showed hyperflexion. In 47 fetuses whose corrected birthweights were 200g or more below the fifth percentile, 10 showed hyperflexion; 3 of these died during labor, and 1 was born severely asphyxiated. The incidence of hyperflexion in small-for-dates fetuses seems to be proportional to the degree of fetal growth retardation, and hyperflexion in grossly small-for-dates fetuses indicates a particularly high risk of severe intrapartum hypoxia. (3 refs.) - *B. J. Grylack.*

Northern General Hospital
Sheffield, England

1968 **HEFFNER, REID R.** Syndrome of absent abdominal muscles: Two cases with microcephaly, polymicrogyria, and cerebellar malformations. *Journal of Neurology, Neurosurgery and Psychiatry,* 33(6):844-850, 1970.

Two cases of the syndrome of absent abdominal muscles with involvement of the central nervous system have been observed. Microcephaly, microencephaly, polymicrogyria, and cerebellar heterotopiae are neuropathological features common to both cases. One case was characterized by the absence of the corpus callosum and partial agenesis of the cerebellar vermis, while the other case

showed a reduction of anterior horn cells at lower thoracic levels. Urological deformities and other musculoskeletal anomalies were also present. The findings in the 2 cases suggest that the syndrome is a developmental defect involving many organ systems and that the influence which produced the defects was present during the first trimester. (45 refs.) - *B. J. Grylack.*

Yale University School of Medicine
New Haven, Connecticut

1969 **VAN GILS, J. F.** Postnatal growth and development in small-for-date babies. In: Stoelinga, G.B.A.; & Van Der Werff Ten Bosch, J. J., eds. *Normal and Abnormal Development of Brain and Behaviour.* Baltimore, The Williams & Wilkins Co., 1971, p. 53-69.

There is little unanimity on the definition of intrauterine growth retardation and its nomenclature. The term "small-for-date" is here used to describe such pathologic retardation. Its causes are maternal and placental. In organ growth, especially the brain's, 3 phases (cell multiplication, cell multiplication and cell-volume increase, and cell-volume increase without cell multiplication) can be seen. Malnutrition during cell multiplication impairs brain myelination and causes growth retardation dependent on the length and severity of the malnutrition. The brains of 9 children dead of malnutrition during the first year of life showed reductions in weight, and lowered DNA, RNA, and protein; the 3 children with the lowest DNA content had a birth weight of less than 2,000 grams. There is varying evidence on the effects of malnutrition on IQ; its effect on brain development depends on the duration of malnutrition in relation to the brain's growth phase. After intrauterine malnutrition, optimal nutrition must be introduced as soon as possible. The degree of brain recuperation after cell-multiplication impairment remains in question. (41 refs.) - *B. Berman.*

1970 **JANS, J.** Non-genetic prenatal influences on psychological development. In: Stoelinga, G.B.A.; & Van Der Werff Ten Bosch, J. J., eds. *Normal and Abnormal Development of Brain and Behaviour.* Baltimore, The Williams & Wilkins Co., 1971, p. 234-243.

There is evidence of significant environmental, non-genetic elements in the prenatal milieu, and it cannot be maintained that a fetus will retain its integrity no matter what environmental exigencies are experienced by the pregnant mother. Although this area of research must overcome methodological problems inherent in the traditional statistical epidemiological approach, there are many studies showing an association between complications during pregnancy and neurological and behavioral disorders. However, a certain association between prenatal factors and postnatal behavior is not adequate evidence of causality. Many investigators have indicated causal connections between maternal emotionality, childbirth complications, and subsequent neonatal behavior. Several researchers have found a positive relation between fetal activity and postnatal development. Sontag reported 8 women who experienced a severe trauma in late pregnancy whose fetuses showed a profound increase in activity level and whose children—although without congenital defects—were generally irritable, hyperactive, and had feeding problems. (38 refs.) - *B. Berman.*

1971 **ELWOOD, J. H.** Anencephalus in the British Isles. *Developmental Medicine and Child Neurology,* 12(5):582-591, 1970.

Regional parity and maternal age differences are not major contributors to the geographical variation in the incidence of anencephalus in the British Isles. Official statistics relating to 3,175,161 live and still births and 6,431 anencephalics which occurred in the British Isles during the years 1965 to 1967 were analyzed. The highest rate, 3.6 per 1,000 total births, was in Northern Ireland. In all areas, more anencephalics were born in winter than in summer. National parity and maternal age differences accounted for less than 5% of the excess of the Irish and Scottish anencephalic rates over England and Wales. Social class may be a factor, but the present data are unable to support this. The results indicate a multifactorial etiology of anencephalus, and the effects of other factors, singly and in combination, need to be studied in relation to this common malformation. (24 refs.) - *S. Glinsky.*

Queen's University of Belfast
Belfast, Northern Ireland

1972 **KALVERBOER, A. F.; LE COULTRE, R.; & CASAER, P.** Implications of congenital ophthalmoplegia for the development of visuo-motor functions (Illustrated by a case with the Moebius syndrome). *Developmental Medicine and Child Neurology*, 12(5):642-654, 1970.

The relationship between visuo-motor disorders and reading and spelling difficulties is not as close as is sometimes assumed. The S was a 12½-year-old boy with a Moebius syndrome who exhibited severely restricted horizontal and vertical eye movements, but no disturbances of gross motor functions. Unusual performances in the visuo-motor tests were shown in the psychological assessment, including a discrepancy between verbal and nonverbal scores in intelligence tests as well as visuo-motor disorders without disturbances in visual discrimination. Comparison of these results with findings in cerebral palsy patients, with data on spatial-constructive development and normative data from children aged 3 to 8 years who were given a Profile and Arrow test, indicates that the S's visuo-motor performances are comparable with those of a normal 4 to 5 year old child. The S's visuo-motor disorders seem to be directly related to his restricted eye movements, yet his reading, writing, and spelling are normal. More cases of congenital ophthalmoplegia must be studied before eye movement restrictions can be definitely related to the development of visuo-motor functioning. (23 refs.) - *S. Glinsky.*

University Hospital
Groningen, The Netherlands

1973 **KOENIG, RONALD H.; & SPIRO, ALFRED J.** Hereditary spastic paraparesis with sensory neuropathy. *Developmental Medicine and Child Neurology*, 12(5):576-581, 1970.

Six members of a family exhibited a neurological disorder characterized by spastic paraparesis associated with sensory abnormalities. The cases were a 17-year-old Negro male, his 47-year-old mother, and 4 siblings: a 15-year-old girl, 14-year-old boy, 7-year-old girl, and 6-year-old boy. Physical findings included presence of foot ulcers in 2 cases, spastic gait with scissoring in 3 cases, hyperflexia in all cases, extensor/extensor response in 6 cases, and sensory abnormalities in 6 cases. The 17-year-old tested as borderline MR, probably due to defensive withdrawal rather than cerebral degeneration. A review of other such rarely occurring cases reveals certain common features, such as gait abnormalities, hyperactive deep tendon reflexes, sensory disturbances, and trophic changes. The mode of inheritance in the present family appears to be autosomal dominant, and the sensory disturbances in this and other cases are but one of the neurological deficits associated with hereditary spastic paraparesis. Lacking pathological confirmation, the nature of the lesion remains speculative. (6 refs.) - *S. Glinsky.*

Albert Einstein College of Medicine
Bronx, New York 10461

1974 **NAEYE, RICHARD L.; MILIC, ANN M. B.; & BLANC, WILLIAM.** Fetal endocrine and renal disorders: Clues to the origin of hydramnios. *American Journal of Obstetrics and Gynecology*, 108(8):1251-1256, 1970.

Newborn anencephalic infants have a subnormal number of nephrons and a reduced cytoplasmic mass in renal tubular cells. The Ss were 31 anencephalic infants who were stillborn or died in the immediate neonatal period and 14 full-term and 6 post-term control infants with normal birth weights. Eight of the anencephalic gestations were associated with hydramnios with increased amniotic volume at birth and had patent collecting tubules, suggesting some fetal micturition. Those without hydramnios had almost closed tubules. The findings are similar in monovular twin partners with the transplacental twin transfusion syndrome. Of 5 additional infants, 2 with isolated absence of the pituitary gland had renal abnormalities like those found in the anencephalic neonates. Two infants with defective anterior lobes of the pituitary had cellular abnormalities centered in the more proximal portions of the renal tubules. (10 refs.) - *S. Glinsky.*

Pennsylvania State University College of Medicine
Hershey, Pennsylvania

1975 **ABRAMOVICH, D. R.** Fetal factors influencing the volume and composition of

liquor amnii. *Journal of Obstetrics and Gynaecology of the British Commonwealth*, 77(10):865-877, 1970.

The role of fetal swallowing and voiding in the control of liquor volume was investigated by measuring liquor volume in 50 pregnancies of between 10 and 20 weeks, by measuring in the dissected fetuses radioactive colloidal gold injected in the amniotic sac about 24 hours before hysterotomy in 10 normal pregnancies of 18-20 weeks gestational age, and by collecting urine from the bladders of 22 fetuses aged 17-22 weeks within 30 minutes of termination of pregnancy. Findings indicate that at 18 weeks gestation the liquor volume increases by 10-13 ml per day. The fetus swallows 4-11 ml per day and may void 7-17 ml per day, while fetal renal function matures with increasing age. Some anencephalic fetuses can swallow considerable volumes of liquor, but the presence or absence of swallowing plays little part in the etiology of polyhydramnios in this condition. A review of clinical evidence together with these findings suggests that, contrary to present theories, fetal swallowing and voiding plays only a small part in the control of liquor volume. There is also a dynamic interchange of water and electrolytes between the feto-placental unit and the mother which undoubtedly plays a role in control of the amniotic fluid, though the exact clinical importance of this exchange remains to be determined. (35 refs.) - *J. C. Moody.*

The Liverpool Clinic
Liverpool L7 7 DE, England

MEDICAL ASPECTS – Etiologic Groupings

Gross brain disease (postnatal)

1976 **ADACHI, MASAZUMI; SCHNECK, LARRY; TORII, JUNZO; & VOLK, BRUNO W.** Histochemical ultrastructural and biochemical studies of a case with leukodystrophy due to congenital deficiency of myelin. *Journal of Neuropathology and Experimental Neurology*, 29(4):601-614, 1970.

A 3½-year-old Jewish boy who had shown arrested psychomotor development since birth was found at autopsy to have a "leukodystrophy." A brain biopsy at age 1 and post mortem examination showed a diffuse absence of myelin sheaths and a marked reduction in oligodendroglial cells. The lack of progression indicates a congenital condition. Histochemical studies showed a decrease in myelin lipids. This entity is not consistent with the globoid, metachromatic, or sudanophilic forms of leukodystrophy and apparently represents a separate genetic entity. (50 refs.) - *E. L. Rowan.*

Kingsbrook Jewish Medical Center
Brooklyn, New York

1977 **RYAN, G. B.; ANDERSON, R. M.; MENKES, J. H.; & DENNETT, X.** Lipofuscin (ceroid) storage disease of the brain, neuropathological and neurochemical studies. *Brain*, 93(3):617-628, 1970.

A boy who died at age 3 yrs 5 mos, after a course indicating severe progressive cerebral degeneration, was found at autopsy to have a lipofuscin storage disease. The wasted brain showed a generalized loss of neurones, and both remaining neurones and macrophages contained large amounts of lipofuscin. This material was also found in histiocytes of the lung, liver, lymph nodes, spleen, thymus, and bone marrow. Chemical examination of the brain tissue showed an increase in glycolipids and cholesterol esters which, when bound by an as yet unidentified substance, appear to make up the lipofuscin material. (32 refs.) - *E. L. Rowan.*

Royal Children's Hospital
Melbourne, Australia

1978 **URBANEK, K.; FARKOVA, H.; & KLAUS, E.** Nishimoto-Takeuchi-Kudo disease: Case report. *Journal of Neurology, Neurosurgery and Psychiatry*, 33(5):671-673, 1970.

A 7-year-old Czech boy exhibited telangiectasia, or "moyamoya disease" symptoms. He experienced paroxysms of unconsciousness and pains in

his extremities, and was found to have a pathological EEG finding and an abnormal angiogram. The causes are probably to be found in circumstances of prenatal development. The angiography with slowing of circulation, an undulated extracranial portion of the internal carotid artery, and tapered stenosis of its supraclinoid segment distinguish "moyamoya disease" from cerebral palsy. The condition is best characterized as a chronic progressive disease with a tendency to advance in attacks which can, however, cease at any stage. It is perhaps etiologically significant that the S's mother, 12-year-old sister, and 3-year-old sister are all of below average intelligence. The general findings in this instance are particularly noteworthy because it involves one of the few non-Japanese cases to be reported in the literature. (9 refs.) - *G. S. Bell.*

University Hospital
Olomouc, Czechoslovakia

1979 **FARIS, AMIN A.; & FLECKENSTEIN, LORAN D.** Subacute necrotizing encephalomyelopathy: Its relationship to central pontine myelinolysis. *Journal of Neurology, Neurosurgery and Psychiatry,* 33(5):667-670, 1970.

A child of 3 years and 11 months showed an increasing inability to walk without assistance, tremor, and a loss of already acquired language ability. A short hospital stay was characterized by excessive somnolence and withdrawal, and death from respiratory embarrassment followed. A diagnosis of subacute necrotizing encephalomyelopathy (SNE) was made on the basis of the usual clinical picture of psychomotor retardation, ataxia, ophthalmoplegia, muscular weakness, and incoordination. Postmortem examination revealed brain lesions, acute bronchial pneumonia and moderate hypertrophy of the right ventricle. It is suggested that SNE and central pontine myelinolysis (CPM) may have a common etiological factor, with the former due to a selective metabolic fault in the oligodendrogliocyte, and the latter, CPM, the result of specific metabolic alterations. (17 refs.) - *G. S. Bell.*

899 Madison Avenue
Memphis, Tennessee 38103

1980 **COOPER, JACK R.; PINCUS, JONATHAN H.; ITOKAWA, YOSHINORI; & PIROS, KATALIN.** Experience with phosphoryl transferase inhibition in subacute necrotizing encephalomyelopathy. *New England Journal of Medicine,* 283(15):793-795, 1970.

Subacute necrotizing encephalomyelopathy (SNE) seems to be the first genetic neurologic disease that results not from an enzyme lack but from the production of an inhibitor that effectively prevents the enzymatic synthesis of a chemical, thiamine triphosphate. The distribution of thiamine, thiamine monophosphate, thiamine triphosphate, and thiamine pyrophosphate in the brain of a child who died of SNE was examined and found to be abnormal: thiamine was extremely high but thiamine triphosphate was deficient. Five cases of SNE and 3 probable cases suggest that the condition is not so rare as previously believed. Further investigation of lesions in other neurologic diseases may disclose similarities with lesions induced by vitamin deficiencies or by the administration of known chemicals. (5 refs.) - *G. S. Bell.*

Yale University School of Medicine
New Haven, Connecticut 06510

1981 **MATSUOKA, TATSUNORI; HAMANAKA, TOSHIHIKO; TAII, SHUNZO; TATEBAYASHI, YOSHINORI; KIJIMA, SHIGEJI; & NISHIKAWA, TADASHI.** Creutzfeldt - Jakob-byo no ichi-akei to shite no a kyusei kaimenjo-no-sho (Subacute spongiform encephalopathy as a subtype of Creutzfeldt-Jakob's disease: Report of two cases). *Psychiatria et Neurologia Japonica,* 72(7):669-690, 1970.

The presence of subacute spongiform encephalopathy (SSE) in the form of spongy type disruptions within the brains of 2 cases who exhibited muscle tone disturbances and died in states of akinetic mutism indicates that SSE is a subtype of Creutzfeldt-Jakob's (CJ) disease. The "periodic synchronous discharges" which were observed in EEG's of each of the 2 cases when they were alive suggest that SSE and subacute sclerosing leucoencephalitis can be diagnosed in part from the appearance of this activity. Case 1 was a 57-year-old male who exhibited symptoms of visual disturbance, parosmia, anxiety, and irritation, 5½ months prior to death. The 1,240g brain was slightly atrophic and revealed typical

spongy lesions as well as fibrous and protoplasmic astrocyte proliferation throughout the cerebral cortex. Case 2 exhibited disturbances of gait 1 year and 9 months prior to death. About a year thereafter, dementia associated with focal signs became prominent and in the later stages disturbances of muscle tone, myoclonic jerkings, and generalized convulsive seizures became prominent, terminating in akinetic mutism. The 1,045g brain was generally atrophic and the spongy state was diffusely developed throughout the cerebral cortex, with senile plaques distributed irregularly. Severe affection of the cerebellum in case 1 was considered unusual and the presence of kuru-like argyrophilic plaques in case 2 was of interest in considering the nosological relationship between kuru and CJ disease. (86-item bibliog.) - *N. Behling.*

Kyoto University
Kyoto, Japan

1982 **AUSTIN, JAMES; SUZUKI, KUNIHIKO; ARMSTRONG, DONALD; BRADY, ROSCOE; BACHHAWAT, BIMAL; SCHLENKER, JANET; & STUMPF, DAVID.** Studies in globoid (Krabbe) leukodystrophy (GLD). V. Controlled enzymic studies in ten human cases. *Archives of Neurology*, 23(6):502-512, 1970.

Enzyme assays were performed at autopsy on 10 children with globoid leukodystrophy (GLD), 18 matched controls, and other controls in an attempt to elucidate the mechanism of increased cerebrosides and decreased sulfatides in GLD. The key enzymatic deviation appears to be a decreased activity of galactocerebroside β-galactosidase in brain, kidney, and liver. (32 refs.) - *E. Kravitz.*

University of Colorado Medical Center
Denver, Colorado

1983 **KATZ, SAMUEL L.; & GRIFFITH, JOHN F.** Slow virus infections. *Hospital Practice*, 6(3):64-74, 1971.

Viral infections with prolonged incubations and slow beginnings of disease processes may be responsible for many chronic and degenerative diseases not yet etiologically explained. Current work derives from that of Sigurdsson and involves research in kuru (possibly a viral meningoencephalitis), first seen in natives of New Guinea, and scrapie (a neurological disease affecting sheep). It is suspected that multiple sclerosis and amyotrophic lateral sclerosis may be slow-virus infections. Studies in transmitting a possible viral agent have been undertaken with kuru, and have produced considerable success with subacute sclerosing panencephalitis (SSPE), a progressive neurologic disorder affecting chiefly school-age children, that is clinically and pathologically strikingly different from most CNS infections. SSPE studies have revealed lesions involving white and grey matter in brain hemispheres; neural-tissue laboratory studies have shown viral cytopathology only in a specific part of the nervous system. Recovery of an agent resembling measles virus seems to confirm the disease's viral origin. Its mystery will be unravelled when a suitable animal model is available. Other systems than the nervous system may be linked to slow viruses, possibly unexplained kidney and liver disorders, and, conceivably, tumorigenesis and aging. (No refs.) - *B. Berman.*

Duke University
Chapel Hill, North Carolina

1984 **KOEHL, R. H.; SOLITARE, G. B.; & HEFFNER, R. R., JR.** Lipomatous hamartoma involving the midbrain and cerebellum of a mentally retarded man. *Journal of Mental Deficiency Research*, 14(Part 3):227-234, 1970.

A 32-year-old Negro male presented with a lipoma of the quadrigeminal plate and cerebellar vermis accompanied by anomalies of adjacent brain structures and other body organs. Lipomata in all portions of the nervous system are generally associated with malformations of adjacent structures. As is usually the case, there was a lack of clinical symptomatology related to the lipoma in this patient. Twelve cases of mental deficiency in association with intracranial lipomata have been recorded previously. The etiology of the retardation is uncertain. History of prematurity in this patient had possible significance, although there was no evidence of anoxic change in the brain autopsy. The corpus callosum was intact, and the cerebral hemispheres were normal. Chromosomal analysis revealed no definite abnormalities. The mental deficiency was probably

related to the concomitant brain anomalies. (28 refs.) - *B. J. Grylack.*

Yale University School of Medicine
New Haven, Connecticut

1985 **MARCHAL, C.; VIDAILHET, M.; VIDAILHET, C.; HUMBEL, R.; GRIGNON, G.; & FLOQUET, J.** La leucodystrophie metachromatique ou sulfatidose chez l'enfant (Metachromatic leukodystrophy or sulfatidosis in children). *Archives Francaises de Pediatrie,* 27(9):996, 1970.

Metachromatic leukodystrophy was diagnosed in 5 cases on the basis of hypersulfatiduria, the presence of renal sulfatides, a sulfatasic deficit in the urine, and, in 2 cases, an excess of sulfatides with a decrease in the cerebrosidic influence on sulfatides at the level of the peripheral nerve. Particularly atypical symtomatology was noted in all cases. A 5-year-old child manifested chronic polyradiculoneuritis with albuminocytologic dissociation. A 3-year-old child presented with tonic convulsive crises accompanied by mental deterioration. A 16-month-old infant showed encephalopathy characterized by the association of mental deterioration with signs of cerebellar and peripheral pyramidal involvement and died after 3 months of the disease course. A 14-month-old infant revealed a clinical picture of pseudomyotonia without other neuropsychic symptoms. Finally, a 14-year-old girl, the sister of the preceding infant, showed encephalopathy with profound MR and spastic quadriplegia. (No refs.) - *B. J. Grylack.*

No address

1986 **SCHULER, D.; SCHONGUT, L.; CSERHATI, E.; SIEGLER, J.; & GACS, G.** Lymphoblastic transformation, chromosome pattern and delayed-type skin reaction in ataxia telangiectasia. *Acta Paediatrica Scandinavica,* 60(1):66-72, 1971.

Analysis of lymphocytic blastic transformation was performed in 6 cases of ataxia telangiectasia to determine whether there was a decrease in such transformation, and the incorporation of ^{3}H-thymidine into nuclear DNA was studied in 3 of these cases. Chromosomal studies were also carried out in 4 cases, and the Candida skin test was performed to examine the delayed-type skin reaction. Results indicated that lymphoblastic transformation was decreased, although there was no correlation between the degree of decrease, skin infections, and plasma IgA level. Incorporation of ^{3}H thymidine showed a decrease of DNA synthesis in ataxic patients. In only one of the subjects did the number of chromosome breakages exceed control values (this subject later developed malignancy), so that increased chromosome breakage cannot be considered a constant feature of this syndrome. (46 refs.) - *N. Jarvis.*

University Medical School
Budapest, Hungary

1987 **SUZUKI, YOSHIYUKI; & SUZUKI, KUNIHIKO.** Krabbe's globoid cell leukodystrophy: Deficiency of galactocerebrosidase in serum, leukocytes, and fibroblasts. *Science,* 171(3966):73-75, 1971.

Serum, leukocytes, and cultured fibroblasts from patients with Krabbe's globoid cell leukodystrophy and from parents of these patients were examined in an attempt to establish a means of antemortem diagnosis of the disease. The activity of galactocerebroside β-galactosidase was markedly diminished in patients with the disease and was lower in the parents of patients than the level of enzyme activity found in normal controls. The activity of the enzyme can be demonstrated only with the use of the specific natural substrate, galactocerebroside, and not with artificial chromogenic substrates. Despite exceptions, the activity of galactocerebroside β-galactosidase is generally lower than normal in parents of patients with Krabbe's disease, as predicted from the assumption that the deficient activity of this enzyme is the genetically determined defect underlying globoid cell leukodystrophy. In the detection of heterozygous carriers, fresh serum provides the best results. Antemortem diagnosis is thus available without organ biopsy. (11 refs.) - *B. J. Grylack.*

University of Pennsylvania School of Medicine
Philadelphia, Pennsylvania 19104

1988 **WIESMANN, U. N.; ROSSI, E. E.; & HERSCHKOWITZ, N. N.** Treatment of

metachromatic leukodystrophy in fibroblasts by enzyme replacement. *New England Journal of Medicine*, 284(12):672-673, 1971. (Letter)

Fibroblasts from 3 patients with juvenile metachromatic leukodystrophy (MLD) and controls were grown for 3 days in a medium to which a preparation of the enzyme arylsulfatase A was added. A decrease in the activity of this enzyme is believed to be the cause of MLD, a genetic disorder characterized by progressive demyelination and MR, with large amounts of cerebroside sulfatide accumulating in the nervous system and in certain organs. The result of enzyme preparation addition showed a change from 2 to 5% up to 50% of normal value for enzyme activity in the cells of Ss. Besides indicating that temporary enzyme replacement in MLD fibroblasts is possible in tissue-culture conditions, the findings revealed repair of the faulty sulfatide degradation. It remains to determine the *in vivo* therapeutic possibilities and to extend these principles to other enzyme deficiencies. (4 refs.) - *B. J. Grylack.*

University of Bern
Bern, Switzerland

1989 **HANEFELD, F.; & CROME, L.** Histochemistry in Krabbe's disease. *Lancet*, 1(7691):195, 1971. (Letter)

The origin of the globoid cells in Krabbe's disease (globoid-cell leukodystrophy) is uncertain. Large amounts of β-glucuronidase in the lysosomes of the globoid cells and of the epithelioid cells surrounding blood vessels have been demonstrated in tissue from one patient, using Hayashi's method of histochemical staining. This finding is in keeping with the view that globoid cells are derived from epithelioid cells. (No refs.) - *J. C. Moody.*

Queen Mary's Hospital for Children
Carshalton, Surrey, England

1990 **STUMPF, D.; & AUSTIN, J.** Qualitative and quantitative differences in sulfatase A which distinguish different forms of classical metachromatic leukodystrophy (MLD). *Journal of Neuropathology and Experimental Neurology*, 30(1):147, 1971.

The various forms of classical metachromatic leukodystrophy (MLD) may reflect enzymic differences. The minute amounts of remaining enzyme activity in MLD were analyzed and four differences emerged. Urinary sulfatase A activity in late infantile MLD was only slightly reduced by a sulfhydryl group inhibitor, while the enzyme in normal controls and in juvenile MLD was inhibited 69 and 75%, respectively. Urinary sulfatase A in juvenile and adult MLD was not inhibited by an excess of its substrate, whereas the normal enzyme is classically inhibited at higher substrate concentrations. In juvenile MLD the sulfatase A was substantially inhibited (68%) by diethyl-dithio-carbamic acid, yet the enzyme in controls was only inhibited 37%. Urinary levels of sulfatase A were quantitatively higher in juvenile MLD than in late infantile MLD. The results suggest that enzymic proteins may differ among the various forms of MLD. (No refs.) - *S. Glinsky.*

No address

1991 **SUZUKI, K.; & SUZUKI, Y.** Krabbe's globoid cell leukodystrophy: Deficiency of galactocerebroside β-galactosidase activity. *Journal of Neuropathology and Experimental Neurology*, 30(1):145, 1971.

The deficiency of galactocerebroside β-galactosidase can explain the morphological and biochemical characteristics of Krabbe's globoid cell leukodystrophy better than the deficiency of sulfotransferase. Using specifically labeled galactocerebroside prepared by oxidation with galactose oxidase and reduction by tritium-labeled sodium borohydride, a profound deficiency of galactocerebroside β-galactosidase in gray and white matter of Krabbe's disease was found. The activity in Krabbe's disease was approximately 5 to 10% of several normal controls and a variety of pathological controls. Results of assay with p-nitrophenyl-β-galactoside as a substrate indicated that galactocerebroside β-galactosidase was a specific enzyme for galactocerebroside. This deficiency was not due to the presence of an inhibitor in the tissue of Krabbe's disease nor to a shift of the pH optima in Krabbe's disease. (No refs.) - *S. Glinsky.*

No address

1992 **LAMPERT, PETER W.; GAJDUSEK, D. CARLTON; & GIBBS, CLARENCE J., JR.** Experimental spongiform encephalopathy (Creutzfeldt-Jakob disease) in chimpanzees. *Journal of Neuropathology and Experimental Neurology*, 30(1):20-32, 1971.

The most significant histopathologic change in the cerebral cortex of chimpanzees affected with Creutzfeldt-Jakob disease is status spongiosus. Electron microscopy studies were carried out on cerebral cortex from 4 chimpanzees, each inoculated with a brain suspension from a different patient with Creutzfeldt-Jakob disease and from 1 chimpanzee injected with a brain suspension from an affected chimpanzee. Rapidly progressive mental and neurological disturbances leading to total incapacitation within 2 months developed in all of the animals 12 to 14 months after inoculation. The encephalopathy was characterized by vacuolar neuronal degeneration, astrocytic proliferation, and a lack of leukocytic infiltration. The status spongiosus corresponded to vacuoles within neuronal projections and to focal clearings and herniations of the cytoplasm of neurons and occasionally also of astrocytes. The vacuoles were bounded by fragmented membranes, proliferated portions of which accumulated at the margins or within the lumen of the vacuoles. Focal disruptions and curling of cell membranes were detected in the wall of swollen neuronal processes. The proliferation of astrocytes was interpreted as a reactive change occurring secondarily to the neuronal damage. The findings show similar histopathologic features to other transmissible spongiform encephalopathies such as kuru, scrapie, and mink encephalopathy. (29 refs.) - *S. Glinsky.*

University of California, San Diego
La Jolla, California 92037

1993 **SUZUKI, K.** Ultrastructural study of experimental globoid cells. *Journal of Neuropathology and Experimental Neurology*, 30(1):145-146, 1971.

Experimentally produced globoid cells in rats contained accumulated cerebroside, and some of the globoid cell inclusions were similar to those found in humans suffering from Krabbe's disease. Purified bovine galactocerebroside was injected into the cerebrum of 12 to 15 day old rats. The animals were perfused with 5% glutaraldehyde in 0.1 M cacodylate buffer at 5, 9, 25, and 35 days post-injection. Proliferative reaction similar to one previously described by Austin and Lehfeldt was seen around the injected cerebroside. The globoid cells were prominent in rats sacrificed in 5 and 9 days, while those sacrificed at 25 and 35 days revealed a lesser degree of reaction. In the electron microscope, the cytoplasm of the globoid cells contained numerous tubular or crystalloid inclusions similar to injected cerebroside. Although some of the inclusions were very similar to those found in globoid cells in humans, their appearance was more variable indicating transformation of cerebroside inside of phagocytic cells. In an earlier stage, the phagocytic cells were intermingled with injected cerebroside, but in later stage they tended to line up with pseudoprocesses toward the cerebroside. Neurons, astrocytes, and myelin sheaths adjacent to the injected cerebroside appeared normal. (1 ref.) - *S. Glinsky.*

No address

1994 **MOOSA, A.** Peripheral neuropathy and ichthyosis in Krabbe's leucodystrophy. *Archives of Disease in Childhood*, 46(245):112-113, 1971.

Krabbe's leukodystrophy with ichthyosis and peripheral neuropathy were present in a 10-month-old female. Development was normal until after the age of 7 months when she lost her ability to sit, became irritable, refused food, and no longer attempted to pull herself to a standing position. At 12 months, she could not sit or stand or take any weight on her legs. She had increased tone around elbow and knee joints and hypotonia around the wrist joints. She had frequent extensor spasms, absence of tendon reflexes, and cold and cyanoid hands and feet. Motor nerve conduction velocities were very slow, confirming the presence of a demyelinating peripheral neuropathy. Central nervous system disturbance was progressive. The patient died at 14 months. Necropsy revealed the characteristic globoid cells of the nervous system found in Krabbe's leukodystrophy. (8 refs.) - *J. K. Wyatt.*

University of Sheffield
Sheffield, England

1995 **STUMPF, DAVID; & AUSTIN, JAMES.** Metachromatic leukodystrophy (MLD). IX. Qualitative and quantitative differences in urinary arylsulfatase A in different forms of MLD. *Archives of Neurology*, 24(2):117-124, 1971.

The enzyme abnormality in late infantile metachromatic leukodystrophy (MLD) is qualitatively different from that in the juvenile form of MLD. The Ss were 7 male and female patients aged 3 years to 63 years, whose age at onset of MLD ranged from 2 years 2 months to 26 years, and a group of controls. Kinetic differences were observed between the urinary sulfatase A in MLD and in controls. In juvenile and adult MLD, the enzyme was not inhibited by an excess of substrate. Normal sulfatase was inhibited by an excess of sulfatase. The MLD patients studied also showed qualitative differences in their responses to p-hydroxi-mercuribenzoate, diethyldithiocarbamic acid, and dl-penicillamine. The data suggest that sulfatase A is abnormal in MLD patients tested and that abnormality of enzymic proteins may differ among the different forms of MLD. (18 refs.) - *S. Glinsky.*

University of Colorado Medical Center
Denver, Colorado

MEDICAL ASPECTS – Etiologic Groupings

Psycho-environmental

1996 **WOLRAICH, MARK; BZOSTEK, BARRY; NEU, RICHARD L.; & GARDNER, LYTT I.** Lack of chromosome aberrations in autism. *New England Journal of Medicine*, 283(22):1231, 1970. (Letter)

Of 25 male and female autistic patients, 5 to 30 years old, only 1 was found to have an abnormal chromosome complement using the micromethod technic. The abnormal karyotype was 44,XXX. This study lends no support to the hypothesis that there may be a specific chromosomal aberration in autistic patients. (2 refs.) - *G. S. Bell.*

Upstate Medical Center
Syracuse, New York

1997 **RITVO, EDWARD R.; ORNITZ, EDWARD M.; TANGUAY, PETER E.; & LEE, JOHN C. M.** *Neurobiological abnormalities in early infantile autism.* 16 p. mimeo. Copies available from author.

Clinical studies of autistic children show common disturbances in developmental rate, perception, motility, relating, and language. This constellation of symptoms is suggestive of disturbed regulation or modulation within the reticular-activating system of the central nervous system. Prolonged periods of observation demonstrate a consistency or stability in the rate of autistic behavior. Such patients had decreased postrotatory nystagmus when tested in the light but not in the dark. Autistic children show an immature EEG sleep pattern and poor regulation of auditory evoked responses during sleep as do normal infants. These signs vanish with growth in normal children. Neurochemical studies show increased blood serotonin and platelet levels in autistic children. All these findings are suggestive of an organic etiology for autism. (27 refs.) - *E. L. Rowan.*

UCLA Center for the Health Sciences
Los Angeles, California 90024

1998 **LEVISON, CATHRYN A.** The development of head banging in a young rhesus monkey. *American Journal of Mental Deficiency*, 75(3):323-328, 1970.

Head banging, a low frequency behavior in nonhuman primates, developed in a rhesus monkey with no previous stereotyped behavior as a result of a particular set of reactions with the experimenter. The S was a male rhesus monkey, born March 11, 1968, separated from its mother at birth and immediately installed in a surrogate rearing apparatus. After one year of deprivation training observation, the monkey developed head banging subsequent to being struck accidentally

on the head during transfer cage training. This response to a negative association was exhibited later in other negative situations. The results suggest that the onset of head banging in infants and MRs may similarly be a consequence of specific, relatively immediate antecedent conditions. (14 refs.) - *S. Glinsky.*

University of Chicago
Chicago, Illinois 60637

1999 SEHAM, MAX. Poverty, illness, and the Negro child. *Pediatrics,* 46(2):305-311, 1970.

Despite rising prosperity among an increasing number of American citizens, there are still many children from poverty environments who are known to suffer from the so-called "deprivation syndrome." This syndrome implies interference with the normal growth and development of the child in the physical, mental, and emotional spheres, frequently as a result of severe social pathology. A disproportionate number of these cases is found among the Negro population. Statistics which consider maternal mortality, prematurity, infant mortality, MR, and physical and emotional disorders reflect the inaccessibility of quality health care to the poor. Project Head Start constituted a governmental move in the right direction. In addition, the general public must respond to obvious needs which have been well documented. (11 refs.) - *B. J. Grylack.*

Group Health Medical Center
St. Paul, Minnesota 55108

2000 EASSON, WILLIAM M. Symptomatic autism in childhood and adolescence. *Pediatrics,* 47(4):717-722, 1971.

A severe perceptual or intellectual handicap such as blindness, deafness, or MR from infancy is the most common cause of childhood autism. Such symptomatic autism is more prevalent than the normal autism of the first few weeks of life, the infantile autism first described by Kanner, or the regressive autism in response to environmental stress. The child with a unique and idiosyncratic perception of the world cannot help but respond differently from "normal" children and have difficulty in adaptation. Early recognition of the underlying problem and appropriate treatment can prevent emotional, intellectual, and social crippling. (12 refs.) - *E. L. Rowan.*

Medical College of Ohio
Toledo, Ohio 43614

2001 LUCAS, ALEXANDER R.; WARNER, KENNETH; & GOTTLIEB, JACQUES S. Biological studies in childhood schizophrenia: Serotonin uptake by platelets. *Biological Psychiatry,* 3(2):123-128, 1971.

In vitro serotonin uptake by platelets was investigated in 16 male schizophrenic patients (mean CA 12.9 years) and 16 hospitalized non-psychotic control patients diagnosed as having personality disorder or brain damage. Medication was withdrawn at least 3 days before the study. The serotonin uptake was determined by the cellophane tube method described by Born and Gillson. There was no significant difference between the mean uptake of 12.43±2.77 picomoles/platelet in the schizophrenics and 12.85±3.04 picomoles/platelet in the controls. Attempts to analyze serotonin uptake with previous drug dosage and duration failed to reveal any important relationship. It is concluded that the *in vitro* serotonin uptake by platelets of schizophrenic children from 9 to 16 years old does not differ from that of hospitalized control Ss. (10 refs.) - *J. C. Moody.*

Lafayette Clinic
Detroit, Michigan

2002 GARMEZY, NORMAN; & NUECHTERLEIN, KEITH. Invulnerable children: The fact and fiction of competence and disadvantage. *American Journal of Orthopsychiatry,* 42(2):328-329, 1972.

While much attention has been given to vulnerable children who due to deprivation are predisposed to psychopathology, invulnerable children, or those deprived children who have resisted disorder, have been relatively ignored in studies. Factors generally predisposing to vulnerability include poor genetic background, disrupted or disorganized family life, pathological rearing conditions, poor parental-child communication, and patterns of early neglect. But little is known regarding invulnerability, where children reared amid deviance and genetic and familial handicap

advance toward maturity, competence, and adaptation. Studies should investigate these invulnerable children and their coping mechanisms, cognitive styles, patterns of work motivation and efficiency, and styles of parenting to understand the basis of competence formation under adverse conditions. (No refs.) - *S. Glinsky.*

University of Minnesota
Minneapolis, Minnesota

2003 **CROUGHS, W.** The influence of emotional deprivation on growth and behavior. In: Stoelinga, G.B.A.; & Van Der Werff Ten Bosch, J. J., eds. *Normal and Abnormal Development of Brain and Behaviour.* Baltimore, The Williams & Wilkins Co., 1971, p. 271-281.

Emotional deprivation among children living with their families is increasing, and the ultimate effects of rejection on physical, emotional, and intellectual development are deleterious. Removal of the mother can result in anaclitic depression, characterized by withdrawal, loss of appetite and weight, dejection, mental and speech retardation, and susceptibility to infection and illness. The syndrome disappears after restoration of the love object. Five percent of infants admitted to Children's Hospital in Pittsburgh in 1966 showed nonorganic failure to thrive, attributed to emotional deprivation. In these infants, emotional factors may alter intestinal motility and absorption, and influence endocrine growth regulation. In 1967, researchers noted a syndrome of emotional deprivation and growth retardation simulating idiopathic hypopituitarism or "deprivation dwarfism." A twin pair (boy and girl) are cited as examples of psychosomatically induced growth failure, manifesting abnormal growth-hormone secretion and adrenal function. After removal from the emotional deprivation of their home, functions returned to normal. (41 refs.) - *B. Berman.*

2004 **RUTTER, MICHAEL; & BARTAK, LAWRENCE.** Causes of infantile autism: Some considerations from recent research. *Journal of Autism and Childhood Schizophrenia,* 1(1):20-32, 1971.

The primary handicap of autism may be a cognitive defect manifested in a severe, extensive deficit in language comprehension, control functions associated with language, and processing of symbolic and sequential information. Social and behavioral abnormalities may develop as secondary consequences. Although the basic cause of the cognitive defect is not known, research data suggest the presence of organic brain disorder. It has not been determined whether autism arises solely because of a cognitive/language defect or whether some interaction with particular personality factors or family environment is necessary. Since there is less evidence for structural brain pathology among intelligent autistic children, it may be that some cases of autism are related to a maturational disorder or a genetically determined condition. (38 refs.) - *J. K. Wyatt.*

Institute of Psychiatry
London, S. E. 5, England

2005 Preemies need cuddling. *Medical World News,* 12(10):38H, 1971.

Sensory deprivation in Isolettes may play a significant role in producing the intellectual impairment, slow learning, motor retardation, hyperactivity, etc., often attributed to neurological defects or minimal brain damage in premature infants. Ten premature babies found to be functioning physically within normal limits 12 hours after birth were arbitrarily assigned to control and experimental groups, with 4 boys and 1 girl in each group. For 10 days the controls were handled only for diaper changes and when being fed or burped. The experimental Ss were additionally fondled for 5 minutes each hour on a 24-hour-a-day basis. The activity of each infant was observed; weight, temperature, startle reflexes, crying time, frequency of elimination, and physical development were recorded daily. The handled infants were significantly more active than those in the control group, and they regained their birth weights in 10.8 days as opposed to 15.4 days for the controls. Follow-up examinations 7 to 8 months after discharge included a complete physical examination and the Bayley Test of Mental and Motor Development. Whereas all of the experimental babies were described as active and physically healthy, 3 of the 5 controls were rated more than 2 standard deviations below the growth mean for their age, and 1 was at the lower limit of normal. Two were suspected of having cerebral palsy, and only 1 was considered to be developing normally. No deleterious effects of the handling process were noted in any of the experimental Ss. (No refs.) - *J. C. Moody.*

MEDICAL ASPECTS – Convulsive Disorders

2006 CHATRIAN, G. E.; LETTICH, E.; MILLER, L. H.; & GREEN, J. R. Pattern-sensitive epilepsy, Part 1. An electrographic study of its mechanisms. *Epilepsia*, 11(2):125-149, 1970.

Four patients with a history of pattern-sensitive epilepsy, one of whom was mildly MR, were subjected to a series of visual patterns under EEG monitoring in order to determine the characteristics of the evocative stimuli. Responses were variable, but parallel lines or stripes subtending 4-41 minutes of arc were the most epileptogenic stimuli. Increasing luminance, greater color contrast and sharpness between lines and background, binocular stimulation, and fixation on the stimulus were directly related to an increase in epileptiform discharges. Such discharges are localized in the posterior part of the head and appear to represent abnormal responses in selected neurons of the visual cortex. (33 refs.) - *E. L. Rowan.*

University of Washington
School of Medicine
Seattle, Washington

2007 CHATRIAN, G. E.; LETTICH, E.; MILLER, L. H.; GREEN, J. R.; & KUPFER, C. Pattern-sensitive epilepsy, Part 2. Clinical changes, tests of responsiveness and motor output, alterations of evoked potentials and therapeutic measures. *Epilepsia*, 11(2):151-162, 1970.

Two of 4 patients with a history of pattern-sensitive epilepsy showed impaired response to visual-motor and audiomotor tests in the presence of pattern-elicited epileptic activity. Both cued responses to visual and auditory stimuli and repetitive motor performance were so affected. Attempts to mitigate pattern sensitivity in one patient by means of spectacles with plus lenses, atropine instillation, and patching one eye were unsuccessful. Individualized schedules of anticonvulsant medications resulted in improvement in the 3 cases where it was indicated, but no patient was seizure-free. (15 refs.) - *E. L. Rowan.*

University of Washington School of Medicine
Seattle, Washington

2008 AICARDI, J.; & CHEVRIE, J. J. Convulsive status epilepticus in infants and children: A study of 239 cases. *Epilepsia*, 11(2):187-197, 1970.

Status epilepticus was observed in 239 children under the age of 15. Generalized seizures and unilateral clonic seizures made up the bulk of the cases. Thirty-seven percent occurred during the first year of life and 73% before age 3. Status was the first ictal manifestation in 77%. Over half the children (57%) had subsequent seizures of the kind associated with organic brain damage. Eleven percent died, 37% had permanent neurological signs, and 48% were MR. Half the cases were symptomatic, equally divided between acute and chronic brain injury, and half were cryptogenic, again equally divided between febrile and nonfebrile cases. Symptomatic status was most common in young male babies and was preceded by other seizures. Early control or prevention of status would appear to be essential. (14 refs.) - *E. L. Rowan.*

Clinique de Paediatrie et Puericulture
Hopital St. Vincent de Paul
Paris, France

2009 FREEMAN, JOHN M. Neonatal seizures – diagnosis and management. *Journal of Pediatrics*, 77(4):701-708, 1970.

Since seizures in the newborn infant are frequently a signal of an underlying disease process, it is important to diagnose and treat the specific underlying cause before irreversible cerebral damage occurs. The forms of neonatal seizures may be varied and subtle; therefore, a high index of

suspicion by the nursery staff is necessary for their recognition. Among the major causes of neonatal seizures are hypoglycemia, hypocalcemia, hypomagnesia, pyridoxine deficiency and dependency, hyponatremia and hypernatremia, meningitis and sepsis, birth trauma and anoxia, and other metabolic disorders. Clues to determine the etiology of a seizure can be obtained from history and physical examination. Infrequent seizures should be treated with phenobarbital administered orally or intramuscularly. Frequent or continuous seizures should be treated with intravenous medications: phenobarbital, paraldehyde, or diazepam. When seizures are not due to a curable metabolic cause, anticonvulsant therapy should be continued for 1 to 2 years. Follow-up studies indicate that initial mortality rate for neonates with seizures is high. However, a large percentage of those who survived the neonatal period were considered normal. (22 refs.) - *J. K. Wyatt.*

The Johns Hopkins Hospital
Baltimore, Maryland 21205

2010 **BIEDER, J.; FAIDHERBE, D.; & HOUILLON, P.** Heterochromie irienne, hypoacousie et epilepsie dans une meme famille (Heterochromia iridis, hypoacusis, and epilepsy in a single family). *Annales Medico-Psychologiques,* 1(1):117-121, 1970.

Infraclinical epilepsy was detected in a male child whose mother suffered from hypoacusis and heterochromia iridis. The heterochromia recurred beyond any doubt in one generation and in the next, where it was associated with hypoacusis. The .epilepsy that was present in the following generation was infraclinical initially and then was connected with language retardation. In a previous study, the association of heterochromia iridis, surdomutism, and epilepsy was noted in a single patient, and systematic examination of deafmutes within the family was recommended. The optimum solution appears to be the systematic investigation of heterochromia iridis in a population of epileptics and in their families. However, the knowledge of this syndrome and its various and dissociated forms will allow for the description of certain infraclinical epilepsies and hypoacuses, as well as important problems in child neuropsychiatry. (8 refs.) - *B. J. Grylack.*

No address

2011 **POUNDS, V. A.** Managing the subnormal epileptic. *Nursing Mirror,* 131(16):38, 1970.

Mental subnormality is frequently accompanied by some form of epilepsy. Although MR children often outgrow seizures, recurrence is always a threat. Brain irritation and behavior patterns will often show at intervals, and behavior disorders must at times be treated as non-functioning epilepsy. Anticonvulsants and other drugs may be required to achieve stability, but constant vigilance is needed to avoid unhealthy side-effects (renal or hepatic disease, skin eruptions, ataxia, and others). Healthy bowel action is imperative, because undue pressure on bowel and bladder nerve endings often triggers fits. All unnecessary emotional stress must be avoided. Above all, the epileptic must not be socially isolated, as this only increases his frustration. Instead, he must be permitted as normal a life as possible. (No refs.) - *B. Berman.*

Selwood Hospital
Frome, Somerset, England

2012 **MERLIS, J. K.** Proposal for an international classification of the epilepsies. *Epilepsia,* 11:114-119, 1970.

A new international classification is proposed for the epilepsies, one that would be useful to all physicians, not merely specialists. Clinical criteria include the seizure form, presence of brain pathology, age, and etiology. EEG criteria emphasize interictal and ictal types. The 3 major divisions are generalized, partial, and unclassifiable epilepsies. Comments are invited by the International League Against Epilepsy. (No refs.) - *E. Kravitz.*

No address

2013 **MISES, J.; LERIQUE-KOECHLIN, A.; & RIMBOT, B.** Post-traumatic epilepsy in children. *Epilepsia,* 11:37-39, 1970.

Brief descriptions are provided of children with post-traumatic epilepsy or EEG abnormalities among 2,500 cases of cranial injury followed for 3 to 15 years. Group 1 contained 21 children with post-traumatic clinical epilepsy. Group 2

had 34 cases manifesting seizures within 10 days after the trauma. Group 3 had 37 cases with no clinical seizures but a typical EEG picture, e.g., spikes or spike-and-wave complexes. (No refs.) - *E. Kravitz.*

Hopital des Enfants Malades
Paris, France

2014 Sectioning the corpus callosum for epilepsy. *Lancet*, 2(7683):1124, 1970. (Editorial)

Section of the corpus callosum and fornix on the side of greatest cerebral damage is suggested in certain cases of intractable and poorly controlled epilepsy in children. This procedure reduces the incidence of seizures without producing significant neurologic deficits in the contralateral half of the body or threat to life. Since the corpus callosum is a major cranial association pathway, sectioning interferes with the integration of sensory input into one hemisphere and motor activity controlled by the other hemisphere. Fortunately, the benefits of sectioning outweigh the undesirable neurologic sequelae. (7 refs.) - *E. Kravitz.*

2015 **ROBOZ, P.; & PITT, D.** Studies on 782 cases of mental deficiency. Part V. *Australian Paediatric Journal*, 6(4):185-191, 1970.

Of 116 patients classified in Heber's Category VII (encephalopathy due to unknown causes, with neurological signs), 4 had an obscure type of progressive neurological disease, 53 had motor disorders, and 54 were epileptic. All cases of progressive neurological disease were SMR and exhibited epilepsy, delayed motor development, and spasticity. Nineteen of the patients with motor disorders were also epileptic, and 64.1% were SMR, 32.1% were moderately MR, and 2.8% were mildly MR. Patients with cerebral palsy had an increased incidence of twins, prematurity, and neonatal symptoms as well as a tendency to a small head. In the 59 cases of epilepsy without cerebral palsy, 7 cases had abnormal electroencephalograms but no overt seizures. There was a raised incidence of prematurity, complications of pregnancy, twins, and psychiatric disorders. In 10 families, there were relatives with epilepsy and/or MR. Psychiatric disorders took the form of hyperactive, explosive behavior or psychotic, autistic, or withdrawn behaviors. (13 refs.) - *J. K. Wyatt.*

Children's Cottages
Melbourne, Victoria, Australia 3101

2016 **GORDON, NEIL.** Folate and vitamin B_{12} in epilepsy. *British Medical Journal*, 3(5716):226, 1970. (Letter)

As indicated previously, the available evidence does not prove that the administration of folic acid to epileptic patients precipitates seizures. It is difficult to draw conclusions from recent uncontrolled studies, and the results of double-blind trials tend to demonstrate that folic acid has no significant effect on behavior or fit frequency. Further trials may prove or disprove the short-term effects on behavior and fit frequency of anti-epileptic drugs which depress serum folate and vitamin B_{12} levels, but the long-term effects must not be ignored. (4 refs.) - *B. J. Grylack.*

Royal Manchester Children's Hospital
Pendlebury, near Manchester, England

2017 **VIUKARI, N.M.A.** Low potassium levels in the cerebrospinal fluid of forty mentally subnormal epileptics. *Journal of Mental Deficiency Research*, 14(Part 3):191-195, 1970.

Forty mentally subnormal epileptics aged 14 to 40 years who were receiving continuous treatment with diphenylhydantoin as the sole anticonvulsant had samples of their cerebrospinal fluid (CSF) measured for various chemical values, with particular attention being paid to potassium. The results showed that all 120 CSF samples from these epileptics revealed potassium levels below the average of 3.3 milliequivalents (meq) per liter (l) as determined elsewhere, and 108 of them were below the 2.96 meq/l average determined in another study. Activation of both glycolysis and cell membrane adenosine triphosphatase (ATPase) by potassium is presumably relatively insufficient to prevent an accidentally excessive sodium influx resulting in a seizure in these Ss. It is important to recognize that neural function depends upon proper concentration and

distribution of electrolytes and that these, in turn, are affected by general body mechanisms for regulation of water balance, pH, and electrolytes. The findings indicate the need for further research on minor potassium and other electrolyte aberrations in epilepsy and in MR. (20 refs.) - *B. J. Grylack.*

University of Helsinki
Helsinki, Finland

2018 **PATRY, GEORGE; LYAGOUBI, SOUAD; & TASSINARI, C. ALBERTO.** Subclinical "electrical status epilepticus" induced by sleep in children. A clinical and electroencephalographic study of six cases. *Archives of Neurology*, 24(3):242-252, 1971.

A subclinical "electrical status epilepticus" was defined and studied in 6 mentally retarded children. The condition consists of a persistent sleep-induced dramatic modification of the electroencephalogram, such as that characteristic of petit mal, which manifests no clinical accompaniment or changes in normal sleep patterns, lasts throughout the night, and subsides upon awakening. The degree of MR was considered directly proportional to the age atoonset of seizures. Speech was lacking entirely in 2 patients, and was severely delayed in 2 others. Although the condition is believed to imply a focal or multifocal brain lesion, the mechanism of extreme activation of epileptic discharges during sleep remains poorly understood. (25 refs.) - *N. Jarvis.*

Hotel-Dieu du Sacre-Coeur
Quebec 8, PQ, Canada

2019 **BILLINGS, DAVID M.; & DEGNAN, MARK.** Kinky hair syndrome. *American Journal of Diseases of Children*, 121(5):447-449, 1971.

A new case of the kinky hair syndrome demonstrates both the already known features and some more recently described roentgenographic findings. The patient was observed between 4½ months and 14½ months and was characterized by white, scanty kinky hair, seizures, psychomotor retardation, and growth failure. Electroretinogram examination revealed responses highly suggestive of blindness. Complete blood cell count, findings of urinalysis, and blood chemistry determinations were within normal limits. Electroencephalograms revealed bilateral spike and wave activity. Microscopic examination of the hair showed pili torti and monilethrix. Roentgenograms of the long bones revealed bilateral metaphyseal spurring mostly at the distal end of the femur. The kinky hair syndrome is now a recognized diagnosable neurodegenerative disorder of infancy with characteristic features. (6 refs.) - *S. Glinsky.*

Department of Pediatrics, New York Hospital
New York, New York 10221

2020 **BAYLIS, MARY E.; CROWLEY, JACQUELINE M.; PREECE, J. M.; SYLVESTER, P. E.; & MARKS, V.** Influence of folic acid on blood-phenytoin levels. *Lancet*, 1(7689):62-64, 1971.

Serum-phenytoin levels fall during folic acid treatment of stabilized epileptic patients. The Ss were 175 male epileptic patients receiving long-term anticonvulsant therapy. Folic acid therapy resulted in a significant fall in plasma-phenytoin levels for folate-deficient patients. In one patient the fall was to below the therapeutic range and was associated with deterioration in fit control. The results suggest that folic acid supplements be given to all patients when they begin anticonvulsant therapy. (15 refs.) - *S. Glinsky.*

Area Laboratory at West Park
Epsom, Surrey, England

2021 **RICHENS, ALAN.** Folic acid in epilepsy. *British Medical Journal*, 1(5740):109, 1971. (Letter)

Trials of folic acid treatment in patients with epilepsy have yielded unsatisfactory results. This has been due to poor design and lack of control in trials, as well as to unreliable measurement of fit frequency and mental state. A recent study in a residential center was unreliable since it overlooked significant increases in type A fits, utilized poor calculation techniques, and excluded patients who had serum phenytoin levels of over 25μg/ml. These patients might have shown the greatest change in fit frequency and mental state. Also it is preferable to select patients for a folic

acid trial using the more reliable criterion of red cell folate to determine chronic folate depletion. Future work should be concentrated on identifying folate-depleted patients more carefully and studying a few in depth, with intensive electroencephalogram studies supplementing records of fits. (6 refs.) - *S. Glinsky.*

St. Bartholomew's Hospital,
London E. C. 1, England

2022 **WIECEK, ZYGMUNT.** Proby Leczenia Properycjazyna (Neuleptilem) Dzieci W "Domu Specjalnym" (Attempts at treatment with Neuleptil in children in a special institute). *Psychiatria Polska,* 5(1):61-63, 1971.

A 20-month clinical trial of Neuleptil showed mixed results in a group of 14 institutionalized boys aged 8 to 15 years. All showed severe MR, epilepsy, hyperactivity, and character disorder. On daily dosage of 30 mg of the drug 10 boys showed markedly improved behavior. Another improved on 60 mg per day, and the rest showed no improvement. The frequency of seizures was not decreased. No side effects were noted. The subjects reverted to prestudy behavior as soon as the drug was withdrawn. (4 refs.) - *E. L. Rowan.*

Spital Psychiatryczny
Ul. Grunwaldzka 3, Lubliniec, Poland

2023 **POND, D. A.** The neuropsychiatry of childhood. *Modern Perspectives in Child Psychiatry,* 1:452-472, 1971.

A discussion of the neuropsychiatric problems of childhood focuses on the special problems of the epileptic. Types of epilepsy and its effects on intelligence and social relationships are especially stressed. The assessment and treatment of brain damage, including cerebral palsy, is also discussed. (34 refs.) - *E. L. Rowan.*

Maudsley Hospital
London, England

2024 **MARSHALL, JOHN.** The classification of the epilepsies. *Developmental Medicine and Child Neurology,* 12(5):660-661, 1970.

The classification of epilepsy has always presented a problem, since our ignorance of the essential nature of the disturbance prevents the use of any classification based solely on etiology. The categories of classification presently rely on the phenomena associated with the disorder, which are drawn from clinical and EEG information, and are not always in accordance with each other. Various recent attempts at classification having thus failed, it is suggested that the epilepsies be classified into 3 parallel categories: the clinical type of seizure, whether generalized, focal, absence, etc.; the type of EEG, interictal or ictal disturbance; and the etiology, where this is known. Such a classification would be of practical value in all aspects of epilepsy management and would facilitate international collection and comparison of data. (1 ref.) - *S. Glinsky.*

National Hospital
London, W.C.1, England

2025 **TAYLOR, DAVID C.** Intelligence, learning and seizures. *Developmental Medicine and Child Neurology,* 12(5):667-668, 1970.

An increasing nterest in the education of epileptic children has focused attention on the possible causal relationship of factors associated with epilepsy and poor intellectual performance in school. One study shows that measured IQ, the obstetric history of the mothers, the early onset of epilepsy, and a temporal locus of EEG foci are related to achievement in epileptics. Other research confirms that most socially handicapped epileptics have in addition some evidence of brain damage, and a family background disturbed by mental illness, social failure, separation, and disharmony. Still other research suggests a continuum of neuropsychological deficit, absent or minimal in children without epilepsy, more obvious in questionable cases, and prominent in epileptic children. Sex differences, task difficulty, and type of pacing are also factors in learning results. Future work must involve measurements of the performance of epileptic children which are much more intimately related to the tasks which the children are called upon to perform in the schoolroom. (12 refs.) - *S. Glinsky.*

Park Hospital for Children
Headington, Oxford, England

2026 **HANSSON, O.** Tryptophan-tolerance test, pyridoxine and epilepsy. *Developmental Medicine and Child Neurology*, 12(5):678-679, 1970. (Letter)

Substantial research seems to cast doubt on the utility of a refinement of the tryptophan-tolerance test as a diagnostic criterion for pyridoxine deficiency. It has been postulated that an increased afterload excretion of certain metabolites is due to decreased activity of kynureninase caused by a pyridoxine deficiency. But using the tryptophan-tolerance test it is not possible to distinguish between a decreased activity of kynureninase and an increased inflow of tryptophan on the kynurenine pathway, that is, increased activity of tryptophan pyrrolase. Also, evidence of decreased kynureninase activity due to pyridoxine deficiency will rarely be found without obvious dietary deficiency or malabsorption, which further limits the efficacy of the tryptophan-tolerance test. Further research in this field should be concentrated on direct determination of enzymes of interest. (9 refs.) - *S. Glinsky.*

University Hospital
Uppsala, Sweden

MEDICAL ASPECTS – Chromosomal

2027 **WEBER, FELICE M.; MULLER, HELGA; & SPARKES, ROBERT S.** Clinical and cytogenetic variations of gonadal dysgenesis in 3 patients. *American Journal of Obstetrics and Gynecology*, 107(7):1092-1098, 1970.

Three patients with gonadal dysgenesis, or Turner's syndrome, show variable clinical and chromosomal patterns. A moderately MR woman with typical features of Turner's syndrome shows a partial deletion of the long arms of one of her X chromosomes (46, XXq-). A second patient with amenorrhea and cleft lip and palate shows a mosaic chromosomal pattern with partial deletions in those cells containing 46 chromosomes (45,X/46,XXp-q-). The third patient is severely retarded and shows a chromosomal mosaicism similar to the second patient. Further clinical and chromosomal descriptions of similar cases may facilitate discovery of the etiology of the aberrations. (15 refs.) - *E. L. Rowan.*

UCLA School of Medicine
Los Angeles, California 90024

2028 **ANDERSON, N. G.; SMITHURST, B.; REGIUS MARY, SR; & WALSH, S.** Trisomy 18. *Lancet*, 2(7682):1085, 1970. (Letter)

Three cases of trisomy 18 confirmed by chromosome analysis were noted among 2,600 deliveries over a recent 1-year period in Australia. This represents an incidence of 1 in 800 live births. (No refs.) - *E. L. Rowan.*

Mater Public Hospital
South Brisbane 4101, Australia

2029 **SALMON, MICHAEL A.; & ASHWORTH, MAUREEN.** Association of autoimmune disorders and sex-chromosome anomalies. *Lancet*, 2(7682):1085-1086, 1970. (Letter)

Two additional cases support the relationship of autoimmune disorders to sex-chromosome anomalies. An 11-year-old girl with rheumatoid arthritis also had an abnormal X chromosome and clinical features consistent with gonadal dysgenesis. A 61-year-old man with scleroderma also had Klinefelter's syndrome. Autoimmune diseases might result from a mutation on the X chromosome. (5 refs.) - *E. L. Rowan.*

Park Hospital for Children
Oxford, England

2030 **DAVIS, RICHARD J.; McGEE, BARBARA J.; EMPSON, JUDITH; & ENGEL, ERIC.** XYY and crime. *Lancet*, 2(7682):1086, 1970. (Letter)

Karyotypes done on 190 male prisoners who were serving life or extremely long sentences at the Tennessee State Prison failed to reveal any with an XYY chromosome complement. The relationship of this complement to criminality will require further unbiased study. (4 refs.) - *E. L. Rowan.*

Vanderbilt University School of Medicine
Nashville, Tennessee 37203

2031 **FRIEDMAN, J. M.; STERNBERG, W. H.; VARELA, M.; & BARCLAY, D. L.** Trisomy-21 in mother and child. *Obstetrics and Gynecology*, 36(5):731-734, 1970.

A 17-year-old Negro female with Down's syndrome gave birth to a premature male infant with striking mongoloid features. Both son and mother had 47,XX,21+ karyotypes. Of 15 similar cases, 7 of the offspring had Down's syndrome and 9 did not. Of the 9 without Down's syndrome, 2 showed clinically different MR. (25 refs.) - *G. S. Bell.*

Tulane University School of Medicine
New Orleans, Louisiana 70112

2032 **HSU, LILLIAN Y. F.; BARCINSKI, MARCELLO; SHAPIRO, LAWRENCE R.; VALDERRAMA, ELSA; GERTNER, MELVIN; & HIRSCHHORN, KURT.** Parental chromosomal aberrations associated with multiple abortions and an abnormal infant. *Obstetrics and Gynecology*, 36(5): 723-730, 1970.

Chromosomal aberrations were found in an infant with phenotypically normal parents. The mother had a pericentric inversion of a Number 2 chromosome and the father was mosaic for trisomy D. The white female infant had bilateral cleft lip and palate, low set ears, asymmetrical chest, a grade II/VI precordial systolic murmur, and simian lines. Chromatid breakage for the 25-year-old mother and 27-year-old father were 10% and 6% respectively in leukocyte culture. An increase of chromosome breaks may have been transmitted from mother to child. (15 refs.) - *G. S. Bell.*

Mount Sinai School of Medicine
New York, New York

2033 **MACAFEE, JEREMY; FORTUNE, DENYS W.; BROWN, JAMES B.; & BEISCHER, NORMAN A.** Fetoplacental function in Down's syndrome (mongolism). *Obstetrics and Gynecology*, 36(5): 735-740, 1970.

In pregnancies associated with Down's syndrome in the fetus, hypertension and preeclampsia were higher than rates for the overall hospital population, indicating that a greater incidence of low estriol excretion would be expected. Five of 10 patients whose fetuses had Down's syndrome had urinary estriol values below normal. Of 100 incidences of mongolism, 9 were malpresentations and 19 were neonatal deaths. (15 refs.) - *G. S. Bell.*

Royal Women's Hospital
Melbourne, Victoria, Australia

2034 **RICHARDS, B. W.** "Renpenning" syndrome. *Lancet*, 2(7671):520, 1970. (Letter)

The publication of an article in 1943 which described sex-linked mental deficiency without proving it casts doubt on the justification of the use of the name "Renpenning syndrome" for sex-linked mental deficiency, since Renpenning and colleagues described their disease 19 years afterwards. Cases described in the interim by 2 other investigators also may have been clinical variants of the same disorder; consequently, the use of descriptive language for this condition may be the best approach at the present time. (4 refs.) - *M. S. Fish.*

St. Lawrence's Hospital
Caterham, Surrey, England

2035 **SHAPIRO, LAWRENCE R.; HSU, LILLIAN Y. F.; & HIRSCHHORN, KURT.** Extra posterior cervical skin: A possible sign of chromosomal aberration in infancy. *Journal of Pediatrics*, 77(4):690-691, 1970.

Chromosomal abnormalities other than trisomy 18, trisomy 21, or translocation mongolism were found in 3 infants with extra posterior cervical skin, but not webbing. Chromosome analysis in

the first case revealed 49 chromosomes with an XXXXY sex chromosome constitution. The second case was interpreted as an example of trisomy 22. Chromosome and radiographic analyses indicated 47 chromosomes with an additional chromosome in the G group (47,XX,G+). Chromosome analysis in a third case revealed XO/XY mosaicism (45,X/46,XY); a buccal smear was 100% chromatin positive. Although excess skin in the posterior cervical region appears to be a nonspecific finding, it may be a valuable sign of possible chromosomal anomaly. (6 refs.) - *J. K. Wyatt.*

Cytogenetics Laboratory
Thiells, New York 10984

2036 **WARREN, RICHARD J.; & RIMOIN, DAVID L.** The G deletion syndromes. *Journal of Pediatrics,* 77(4):658-663, 1970.

Deletion of G chromosomes can result in at least 2 distinct clinical syndromes: G deletion syndrome I (antimongolism) and G deletion syndrome II. MR is the only symptom consistently found in both syndromes. Consistent phenotypic characteristics of G deletion syndrome I include antimongoloid slants, hypertonia, prominent nasal bridge, micrognathia, growth retardation, and nonspecific skeletal malformation. Cases of G deletion syndrome II present epicanthal folds, hypotonia, and syndactyly of the second and third toes. Although clear clinical delineations have been found between the 2 syndromes, no specific karyotypic or dermatoglyphic differences are apparent. (30 refs.) - *J. K. Wyatt.*

Washington University School of Medicine
St. Louis, Missouri

2037 **RICCARDI, VINCENT M.; ATKINS, LEONARD; & HOLMES, LEWIS B.** Absent patellae, mild mental retardation, skeletal and genitourinary anomalies, and C group autosomal mosaicism. *Journal of Pediatrics,* 77(4):664-672, 1970.

A unique combination of phenotypic features presented by 3 unrelated children (2 boys, 1 girl) appears to characterize a new syndrome. Two patients (1 boy, 1 girl) presented mosaicism for an extra submetacentric autosome comparable in size to a C group (6-X-12) chromosome. The third patient was mosaic for a large acrocentric autosome. Common physical features in all cases included: height and weight below the tenth percentile; unusual, similar facies; abnormal, similar pinnae; strabismus; abnormal foot structure; fibular/plantar toe curvature; absent patellae; genitourinary anomalies; MR; and unusual dermatoglyphics. Physical features common to any 2 patients were: 6 lumbar vertebrae; spina bifida; limited elbow supination; palmar tendon shortening; incomplete thumb abduction; short, otherwise normal nails; abnormal sternum; conductive hearing loss; clumsiness; and small optical discs. The MR observed in 2 patients was characterized by poor language development, poor visual-motor coordination, and limited abstraction abilities; however, they exhibited good ability in reading, arithmetic, and other rote tasks. (4 refs.) - *J. K. Wyatt.*

Massachusetts General Hospital
Boston, Massachusetts 02114

2038 **COLEMAN, RICHARD.** Klinefelter's syndrome: A review of the research. *Journal of Mental Subnormality,* 16(31):103-105, 1970.

Since the original description of Klinefelter's syndrome in 1942, consensus in research with regard to symptomatology, incidence, or correlation with MR has not been achieved. There has been general agreement that the highest frequencies occur at the upper limits of MR. Among the clinical features described as etiologically related to Klinefelter's syndrome are diabetes mellitus, maternal age, genetic factors, and malignant neoplasm. The results of histological and karyotyping studies have been equivocal. Additional research aimed at identifying specific symptomatology is needed. (24 refs.) - *J. K. Wyatt.*

University of North Carolina
Chapel Hill, North Carolina

2039 **BROGGER, ANTON.** Anticonvulsant drugs and chromosomes. *Lancet,* 2(7680):979, 1970.

Repeated cultures of leukocytes from an MR boy receiving 500 mg of the anticonvulsant ethotoin

daily for 8 months, revealed that 36.2% of the cells had chromosome and chromatid breaks, an unusually high frequency. The preponderance of the chromosomal anomalies occurred in chromosome 3. The *in vitro* effect of ethotoin on chromosomes was investigated. Thirteen cultures were treated for 12 to 24 hours with doses of from 0.5 to 500 mg/ml of the drug. As a result, 5 to 18% of the leukocyte metaphases were damaged with the controls at 10 to 16%. It was concluded that ethotoin did not induce chromosome aberrations *in vitro*. (No refs.) - *B. J. Grylack.*

Norsk Hydro's Institute for Cancer Research
Oslo 3, Norway

2040 **REED, TERRY E.; BORGAONKAR, DIGAMBER S.; CONNEALLY, P. MICHAEL; YU, PAO-LO; NANCE, WALTER E.; & *CHRISTIAN, JOE C.** Dermatoglyphic nomogram for the diagnosis of Down's syndrome. *Journal of Pediatrics*, 77(6):1024-1032, 1970.

Dermatoglyphics from 32 pattern areas were compared in 250 patients with Down's syndrome (proven by karyotype) and in 332 normal control subjects, and 4 patterns were found to account for most of the variance between these groups. Adjusted discriminant scores were then computed for the right hallucal pattern, right and left index finger patterns, and right *atd* angle. The nomogram derived from these 4 scores identified 81% of proven cases of Down's syndrome and will be of value in suggesting this diagnosis in doubtful cases and confirming it without the need for karyotype in clinically obvious cases. (19 refs.) - *E. L. Rowan.*

*Indiana University Medical Center
Indianapolis, Indiana 46202

2041 **BREG, W. ROY; STEELE, MARK W.; MILLER, ORLANDO J.; WARBURTON, DOROTHY; DeCAPOA, ADRIANA; & ALLDERDICE, PENELOPE W.** The cri du chat syndrome in adolescents and adults: Clinical finding in 13 older patients with partial deletion of the short arm of chromosome No. 5(5p-). *Journal of Pediatrics*, 77(5):782-791, 1970.

Thirteen patients aged 12 to 55 with the cri du chat syndrome (partial deletion of the short arm of the Number 5 chromosome) show clinical features different from infants with this syndrome. These adolescents and adults showed severe MR, microcephaly, dental malocclusion, short stature, thin face, divergent strabismus, short philtrum, and short metacarpals and metatarsals. General health was good. This condition is not a rare cause of severe MR. (31 refs.) - *E. L. Rowan.*

Southbury Training School
Southbury, Connecticut 06488

2042 **MILLER, ORLANDO J.; BREG, W. ROY; WARBURTON, DOROTHY; MILLER, DOROTHY A.; DeCAPOA, ADRIANA; ALLDERDICE, PENELOPE W.; DAVIS, JESSICA; KLINGER, HAROLD P.; McGILVRAY, EVE; & ALLEN, FRED H.** Partial deletion of the short arm of chromosome No. 4(4p-): Clinical studies in five unrelated patients. *Journal of Pediatrics*, 77(5):792-801, 1970.

Five patients with 4p- syndrome (deletion of short arm of late replicating B chromosome) show characteristics which distinguish them from patients with the cri du chat syndrome (5p-). The catlike cry is absent and these infants are smaller and more profoundly MR. Characteristic facies (beaked nose and "fish-mouth"), genital anomalies, preauricular and sacral dimples, seizures, eye anomalies, delayed bone growth, and dermatoglyphic characteristics (hypoplastic dermal ridges and low ridge counts) are noted. The parents of children in this series had poor reproductive histories. (24 refs.) - *E. L. Rowan.*

Columbia University
New York, New York 10032

2043 **BELMONTE, M. M.; COLLE, E.; MURPHY, D. A.; & WIGLESWORTH, F. W.** Nonketotic hyperosmolar diabetic coma in Down's syndrome. *Journal of Pediatrics*, 77(5):879-881, 1970.

A 28-month-old boy with Down's syndrome presented with a nonketotic, hyperosmolar, hyperglycemic coma. This is only the tenth such coma

reported in a child and the fourth with Down's syndrome. The boy subsequently developed bilateral femoral artery thromboses. Despite the inauguration of adequate therapy, the boy died. The exact cause of death was uncertain. (5 refs.) - *E. L. Rowan.*

McGill University
Montreal, Canada

2044 TAKEI, HIROSHI. Seishinshojo o yusuru Klinefelter-shojogun no noha to testosterone chiryo no koka (EEG of Klinefelter's syndrome with psychiatric disorders, and the effect of testosterone treatment). *Psychiatria et Neurologia Japonica*, 72(10):983-1021, 1970.

Testosterone and human chorionic gonadotropin (HCG) were separately administered to patients with Klinefelter's syndrome associated with psychiatric disorders. Testosterone effected a significant improvement in their EEG abnormalities and clinical symptoms. EEG examination was made of 20 patients, aged between 11 and 68 years, with Klinefelter's syndrome identified by chromosomal analysis of peripheral lymphocytes. Most of the cases showed irregular and slow alpha activities at rest. Alpha blocking response was incomplete in 10 cases, and in 3 of them paradoxical alpha blocking response was observed. In most cases the slow activities were bilaterally synchronous, consisting of random but frequent theta waves, transient theta waves, or theta bursts. Testosterone was administered to 6 cases, and successive EEG recordings were made of them. Three showed a decrease in abnormal slow activities, and 1 showed a decrease in both abnormal slow and fast activities. In another case, persistent drowsiness and alpha activity appeared. In these 5 cases regular basic alpha activity around 10 cycles per second appeared and the EEG pattern improved markedly. One case was unchanged during testosterone administration. Marked clinical effects of testosterone were observed on the endocrine psychosyndrome of M. Bleuler, such as diminished drive and unstable emotion, and on hypochondriac complaints. HCG was administered to 2 cases. EEG changes caused by HCG were an increase in slow activities, accompanied by more irregular basic activities or by fast activities. In 1 psychotic case HCG appeared to have caused a relapse of psychotic symptoms. (66-item bibliog.) - *N. Behling.*

Tokyo University
Tokyo, Japan

2045 ZANG, K. D.; & ZANKL, H. Chromosome size and hypodiploidy. *Nature*, 228(5273):778-779, 1970. (Letter)

The numerical loss of chromosomes noted in a study on 4,884 mitoses was largely due to mechanical factors, not only chromosome size. Both metaphases with 45 chromosomes and mitoses which lost up to 6 chromosomes were studied. The hypodiploidy rate was 12.9% (Neurath, 16.3%). The source material was blood lymphocytes from healthy persons, including mothers of mongoloid children. The highest loss rate was in the metacentric E chromosome, and the lowest rate was in the nucleolus-organizing chromosomes (O and G). (10 refs.) - *E. Kravitz.*

Max Planck Institute for Psychiatry
Munich, Germany

2046 BROCK, D.J.H.; & SINGER, J. D. Red-cell triosephosphate isomerase and chromosome 5. *Lancet*, 2(7683):1136-1137, 1970. (Letter)

Evidence is discussed which indicates that localization of the red-cell triosephosphate isomerase (TPI) gene on the short arm of chromosome 5 is unlikely. Apparent anomalous findings compared with the work of Sparkes are explained as due to differences in substrate concentrations. The use of saturating levels of substrate is recommended; the assay system of Sparkes used lower concentrations of substrate. Under the latter conditions, enzyme activity is a function of substrate level as well as enzyme quantity. Thus, erroneous half-normal values can easily be obtained instead of normal values, e.g., in cri-du-chat patients. (3 refs.) - *E. Kravitz.*

Western General Hospital
Edinburgh EH4 2HU, Scotland

2047 LUNDBERG, P. O.; & WAHLSTROM, J. Hormone levels in men with extra Y chromosomes. *Lancet*, 2(7683):1133, 1970. (Letter)

Plasma growth hormone levels were normal in 4 adult males with supernumerary Y chromosomes.

Three subjects were 47,XYY and one was 48,XXYY; all were over 6 feet tall, hospitalized, MR patients. Thus, the above-average height of subjects with supernumerary Y chromosomes could not be explained by the presence of high levels of growth hormone in these adults. Such hormone determinations in non-adults might present a different picture. (4 refs.) - *E. Kravitz.*

Academic Hospital
Uppsala, Sweden

2048 LEHRNBECHER, WOLFGANG; LUCAS, GEORGE J.; PICCIANO, DANTE; & JACOBSON, CECIL B. Progressive neuromuscular and skeletal disorders in a male with an acrocentric supernumerary chromosome. *Nature*, 227(5258):612-613, 1970. (Letter)

The case of an elderly hospitalized male with an acrocentric supernumerary chromosome is described. There was an increasing generalized muscle weakness. Neurologic defects began with extraordinary hypotonia of the entire body. There was no myotonic response in any muscle. There was rapid mental deterioration. A modal chromosomal number of 47 was noted; there was an extra subterminal chromosome larger than the G autosomes. There were indications of changes in the amino acid sequence of collagen. Autoradiographic data were similar to those for most XYY persons, but the possibility of a deleted fragment is also suggested. (29 refs.) - *E. Kravitz.*

Overholser Division of Clinical Research, NIMH
Washington, D. C.

2049 POLANI, P. E.; ANGELL, ROSLYN; GIANNELLI, F.; DE LA CHAPELLE, A.; RACE, R. R.; & SANGER, RUTH. Evidence that the Xg locus is inactivated in structurally abnormal X chromosomes. *Nature*, 227(5258):613-616, 1970. (Letter)

Data from 19 female patients are presented in support of the hypothesis that the Xg locus is inactivated in structurally abnormal X chromosomes. Chromosomes from 4 of these cases were studied directly. In one group of patients, the short arm of the X chromosome was missing, and there were isochromosomes for the long arm of the X or other short-arm deletions. In the second group, there were long-arm deletions or short-arm isochromosomes (the long arm is missing). Thus, it is suggested that the Xg^a allele at the Xg locus, known to be on the X chromosome, does not show its presence when on a structurally abnormal X chromosome, irrespective of whether long or short arms are missing. (18 refs.) - *E. Kravitz.*

Guy's Hospital Medical School
London SE1, England

2050 GEORGE, K. P.; & POLANI, P. E. Y heterochromatin and XX males. *Nature*, 228(5277):1215-1216, 1970. (Letter)

Studies of 2 males, both 46,XX, as in a normal female, with no indication of sex-chromosome mosaicism, revealed that Y heterochromatin was not present. One case, age 16 years, was chromatin-positive with gynecomastia and an undescended testis that was Klinefelter-like on histologic examination. The other case, age 7 months, was also chromatin-positive. Photomicroscope observations of cells from these subjects suggest that the presence of the Y long-arm heterochromatin is not absolutely necessary for male sex determination. (14 refs.) - *E. Kravitz.*

Guy's Hospital Medical School
London SE1, England

2051 GOODMAN, STEPHEN I.; & BROWDER, J. ALBERT. Hydroxylysinuria in association with trisomy 21. *Lancet*, 2(7683):1141-1142, 1970. (Letter)

A case of hydroxylysinuria in a 5-year-old girl with trisomy 21 is described. The serum free-hydroxylysine was 0.31 mg/100 ml (none is detectable normally). The ratio of free to peptide-bound hydroxylysine in the patient's urine suggests that the free hydroxylysinuria comes from a mutation of a protein which degrades free hydroxylysine. The presence of trisomy may help to explain the MR associated with hydroxylysinuria. (7 refs.) - *E. Kravitz.*

University of Colorado Medical Center
Denver, Colorado 80220

2052 STERN, J. Biochemistry of Down's syndrome. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 143-157.

Although no biochemical abnormality has been found that is both pronounced and present in every case, Down's syndrome victims do as a group manifest biochemical differences from normals and from other retardates. Because of many uncontrolled factors, a biochemical abnormality should not be considered characteristic of the disease unless it is confirmed in several centers, preferably in more than one country. One of the earliest findings in Down's syndrome was an increase in gamma globulin and a decrease in albumin. Abnormalities have been noted at various levels of the nicotinamide and serotonin pathways of tryptophan metabolism. Investigations were made of more than 24 white- and red-cell enzymes to detect a gene-dosage effect due to the extra chromosome 21, but, although deviations were found in more than half, no simple gene-dosage effect was noted. Down's syndrome patients show significantly higher uric-acid levels than other retardates, and urinary uric acid rises with elevated white-cell turnover and declines with renal insufficiency. Some workers have noted a high incidence of protein-losing enteropathy. There is an increased incidence of diabetes in Down's syndrome, as well as an increased frequency of thyroid antibodies. Little is known about the biochemistry of brain changes in Down's syndrome; diffuse and variable, they are sufficiently severe to account for the MR universally present in this disease. (70 refs.) - *B. Berman.*

Queen Mary's Hospital for Children
Carshalton, Surrey, England

2053 BENSON, P. F. RNA synthesis by Down's syndrome leucocytes. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 161-167.

Studies of the synthesis rates for different RNA types, carried out on cultured lymphocytes with or without phytohemagglutinin stimulation, demonstrated that: (1) incorporation rates of ^{3}H-uridine into RNA were higher in Down's syndrome lymphocytes than in controls; (2) labeling of ribosomal RNA was more rapid in Down's; and (3) radioactivity of RNA with polydisperse sedimentation was higher in Down's. The increased incorporation of ^{3}H-uridine could result from an accelerated chromosomal transcription rate. (6 refs.) - *B. Berman.*

Guy's Hospital Medical School
London, England

2054 YIELDING, K. LEMONE. The effect of chromosome redundancy on gene expression. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 173-182.

Much information is needed about the gene products involved in predicting results of chromosomal redundancy. In addition, one must know the extent and mechanisms of gene control, whether steady-state or pre-steady-state conditions control the system, and whether the overall cell economy involves rate-limiting processes. Predictions that chromosomal redundancy may yield increased, unchanged, or depressed expression and may come at steady state can be extrapolated from known model systems and regulatory mechanisms. Although such predictions are still difficult, clarification of developmental abnormalities may well come from continued study of gene expression as a function of gene dose; such study may also clarify gene expression in higher life forms. (23 refs.) - *B. Berman.*

University of Alabama Medical Center
Birmingham, Alabama

2055 REES, H.; & JONES, R. N. Chromosome gain in higher plants. In: Benson, P. F., ed. *Cellular Organelles and Membranes in Mental Retardation.* Edinburgh and London, Churchill Livingstone, 1971, p. 185-206.

Plants have a remarkable capacity for quantitative change in chromosome material. These changes, which have physiologic and genetic consequences, fall into 4 categories: (1) those involving whole sets of chromosomes (polyploidy); (2) the addition of whole chromosomes,

but not complete sets (aneuploidy); (3) changes involving new, supernumerary, or β chromosomes; and (4) change involving parts of chromosomes, including the amplification of localized segments. Polyploid plants show different features from diploids, and have no problems in the efficiency of mitotic divisions; in meiosis, there is incompetence and sometimes infertility. Despite the hazards, polyploidy has contributed to the evolution of higher plants. Aneuploidy, also, is more frequent in plants than in higher animals, deriving from irregular chromosome disjunction at meiosis in polyploids. Some 200 species of flowering plants show β-chromosomes, which produce unusual but adaptive phenotype variations. They represent a gain in nuclear genetic material, and their excess is explained by their non-Mendelian inheritance. Overall, however, their physiologic effects are deleterious, even though they confer some adaptive benefits. Widespread among flowering plants is gain or amplification of smaller segments within chromosomes; since it accounts for much of DNA variation among species, it affects the mitotic cycle, whose duration correlates with DNA amount. (48 refs.) - *B. Berman.*

University College of Wales
Aberystwyth, Wales

2056 KARDON, NATALINE; HSU, LILLIAN Y.; BERATIS, NICHOLAS; & HIRSCHHORN, KURT. Trisomy 18. *Lancet,* 2(7676):782, 1970. (Letter)

A significant increase in the occurrence of trisomy 18 was noted on a newborn service over a 6-month period. An incidence of 5 cases confirmed by chromosomal analysis out of approximately 2,500 deliveries represented a 7- to 8-fold increase over the frequency in the general population. The maternal ages as a whole were not remarkable. Australia antigen and antithyroid antibodies are being investigated for their association with possible etiological factors. (3 refs.) - *B. J. Grylack.*

Mount Sinai School of Medicine
New York, New York

2057 DIASIO, ROBERT B.; & GLASS, ROBERT H. The Y chromosome in sperm of an XYY male. *Lancet,* 2(7686): 1318-1319, 1970. (Letter)

The sperm of a known XYY man was examined in an attempt to evaluate the transmission of chromosomes from the XYY man to his offspring and to explain the sex ratio of these progeny. A quinacrine-dihydrochloride stain was used to stain the distal end of the long arm of the Y chromosome. Those spermatozoa containing fluorescent body (F-body) within the head were thought to represent Y-bearing spermatozoa. One thousand spermatozoa were counted. Of these, approximately 70% contained 1 F-body, 5% contained 2 F-bodies, and the rest contained no obvious F-bodies. Since no technique is available for selective staining of the X chromosome in sperm, the percentage carrying the XY chromosomal complement cannot be determined with any certainty. The finding of a small but significant number of sperm possessing 2 F-bodies does not appear to be consistent with the fact that only 1 case in which both father and son had an XYY chromosomal constitution has been reported. (14 refs.) - *B. J. Grylack.*

Yale University School of Medicine
New Haven, Connecticut

2058 JONGBLOET, P. H. Chromosomal aberrations and month of birth. *Lancet,* 2(7686):1317-1318, 1970. (Letter)

Syndromes in children resultant from chromosomal aberrations occur with greatest frequency in the spring and autumn. These fluctuations are synchronous with those of the birth curve of normal children and thus are linked to specific seasons, although with a greater amplitude. Theoretical consideration of these changes suggests that the disproportionate increase of pathological conceptions during the spring and autumn and the disproportionate decrease in the winter and summer are caused by the interference of basic estric patterns which form the basis of the normal conception curve. Such interference could slow the preovulatory or intrafollicular ripening of the egg-cell, resulting in overripeness-ovopathy, with meiotic nondisjunction as a consequence. Analogous phenomena in spermatogenesis through the influence of climatological factors cannot be excluded. (4 refs.) - *B. J. Grylack.*

Observation and Treatment Centre
for Mentally Retarded
Ottersum, The Netherlands

2059 **CARTER, C. H.; SMITH, G. F.; & SCHINDELER, JOAN.** Choanal atresia and bifid thumb associated with an abnormal D group chromosome. *Journal of Mental Deficiency Research*, 14(Part 3):221-226, 1970.

History and physical examination of a male child aged 20 months revealed low birth weight, wide set eyes, broad base of nose, epicanthal folds, choanal atresia, large low set ears, micrognathia, congenital heart disease, genital abnormalities, puffiness and creasing of skin of the upper limbs, broad bifid terminal phalanx of thumb, clubbed feet, and delayed physical and mental development. Karyotype analysis showed a missing chromosome in the D group and an extra chromosome in the C group. When pairs of chromosomes were matched according to size and centromere location, the long arms of the extra C group chromosome matched the long arms of the missing D group chromosome, and the short arms of the D group chromosome contained extra chromosomal material. Since neither parents nor an MR brother of the S showed an enlargement of the short arm of a D group chromosome, it appears that the extra translocation of the chromosomal segment probably occurred during meiosis as a *de novo* event. (15 refs.) - *B. J. Grylack.*

Sunland Training Center
Orlando, Florida

2060 **FABIA, JACQUELINE; & DROLETTE, MARGARET.** Life tables up to age 10 for mongols with and without congenital heart defect. *Journal of Mental Deficiency Research*, 14(Part 3):235-242, 1970.

Sex-specific life tables with categorization regarding presence or absence of congenital heart disease have been constructed for 2,421 mongols born alive to Massachusetts residents from 1950 through 1966. In this series, 29.9% of 1,180 girls and 27.2% of 1,241 boys had congenital heart disease. Of all the mongols, 67.2% of boys and 61.9% of girls were alive at age 10. Of the mongols with congenital heart disease only 44.9% of boys and 32.0% of girls survived the first 10 years of life. The one finding which is difficult to explain is that fewer females survived than males among mongols with congenital heart disease. It is possible that trisomy G in general may be more lethal to girls, in conjunction with the fact that the heart malformation may be diagnosed and reported more readily when the child dies. Another possibility is that female mongols may have different types of cardiac defects, a consideration which could not be investigated in this series. (19 refs.) - *B. J. Grylack.*

Harvard University
Boston, Massachusetts 02115

2061 **LAXOVA, RENATA; & KAMARYT, J.** Amylase heterogeneity in patients with Down's syndrome. *Journal of Mental Deficiency Research*, 14(Part 3):250-253, 1970.

Random urine specimens were obtained from 297 patients with standard trisomy G Down's syndrome, 6 patients with translocation Down's syndrome, and 270 healthy controls in order to study the phenotypical variants and gene frequencies of amylase heterogeneity, a recently reported polymorphism. Results revealed no significant differences in the frequencies of the phenotypes in the 3 groups studied. The findings help to exclude the possibility that the additional G group chromosome present in Down's syndrome is the site of either of the loci controlling amylase heterogeneity. One locus is known to control the salivary amylase fraction, and the other regulates amylolytic activity originating in the pancreas. (12 refs.) - *B. J. Grylack.*

Harperbury Hospital
Hertfordshire, England

2062 **YANAGISAWA, S.; & SHUTO, T.** Sex chromatin survey among mentally retarded children in Japan. *Journal of Mental Deficiency Research*, 14(Part 3):254-262, 1970.

A sex chromatin survey of 1,373 male and 881 female institutionalized mental defectives and 791 male and 638 female children in special classes for MRs was performed to determine the frequency of sex chromatin abnormalities in a large group of MR children. Buccal mucosal cells were obtained for sex chromatin screening tests, and chromosome analyses were performed on

lymphocytic cultures from peripheral blood. The IQ distribution of institutionalized mental defectives was lower than that of pupils in special classes for MR children. Of the institutionalized mental defectives, 0.58% of males were chromatin positive and 0.79% of females had sex chromatin abnormalities. In the special classes, 0.83% of males and 0.79% of females had sex chromatin abnormalities. The mean IQ of XXY males (60.4±32.0) was higher than that of XXX females (46.3±11.9), with the males being found generally among EMR groups in special classes. (43 refs.) - *B. J. Grylack.*

Yamaguchi University School of Medicine
Yamaguchi, Japan

2063 HAMBERT, G.; & OLANDERS, S. Prevalence of excess Barr bodies in Swedish institutions for the mentally handicapped. *Journal of Mental Deficiency Research,* 14(Part 3):263-265, 1970.

Buccal smears of 778 males and 651 females, representing all of the institutionalized MRs in the 3 western counties of Sweden, were examined in order to determine the prevalence of excess Barr bodies among this population. When an IQ of 70 was used as a dividing line, 650 of the examined females were found normal for sex chromatin, and 1 was negative. Six out of 778 males, or 0.77%, were chromatin positive. This rate of positive sex chromatin found in males agrees well with figures from other countries. A greater prevalence of positive sex chromatin found for the antisocial retarded as compared with other retarded in a previous study indicates a positive correlation between a supernumerary X chromosome and difficulties in social adjustment. (5 refs.) - *B. J. Grylack.*

St. Jorgen's Hospital
Hisings Backa, Sweden

2064 FENICHEL, GERALD M. The neurological disorders of children. *Clinical Proceedings Children's Hospital,* 27(3):67-73, 1971.

Although genetic defects as a cause of MR have received major scientific attention, they represent the numerically smallest nosological entity in regard to either static or neurological impairment. The single largest identifiable cause of MR is chromosomal abnormalities, especially trisomy 21. These abnormalities may be caused by radiation, physical factors, viruses, and drugs. Viruses produce 3 types of alterations in the chromosome: breaks, fragmentations, and spindle disorganization. Meningitis and encephalitis account for at least 40% of MR in which brain damage occurred postnatally. Other infectious diseases which can cause neurological disorders include mumps and measles. Intrauterine infection is the single largest cause of chronic brain syndromes, including MR. Recent experiments have established that viruses pass through the placenta and that fetal damage is not necessarily related to degree of maternal illness. Viral damage to the fetus can be direct or indirect. (No refs.) - *J. K. Wyatt.*

Vanderbilt University School of Medicine
Nashville, Tennessee

2065 RICHARDS, B. W. Observations on mosaic parents of mongol propositi. *Journal of Mental Deficiency Research,* 14(4):342-346, 1970.

An analysis of maternal age at birth of mongoloid child, grandmaternal age at birth of mosaic parents, and proportion of trisomic cells was made of 11 parents of mongoloid offspring collected from the medical literature. Parents exhibited 46/47 mosaicism without structural change and had a mean maternal age at the birth of the mongoloid child of 27.5 years. Mean maternal age at birth of mosaic mongoloid children was appreciably lower (31.9 yrs) than mean maternal age at birth of trisomic mongoloid children (33.3 yrs). Mean grandmaternal age at birth of mosaic parents was 33.7 years. Although these findings suggest that the majority of mosaic parents arose from trisomic zygotes, 10 of 11 of the maternal mosaics presented less than 34% trisomic cells in cultured leukocytes. Since these mothers did not exhibit the reduced fertility associated with Down's syndrome, they may represent a selection of mosaics who had undergone a reduction in the proportion of trisomic cells during embryogenesis or after birth. (35 refs.) - *J. K. Wyatt.*

St. Lawrence's Hospital
Caterham, Surrey, England

2066 **ZAJACZKOWSKA, KRYSTYNA; KORNISZEWSKI, L.; & WOLFF-PLODOWSKA, ANNA.** A case of quintuple-X syndrome (49,XXXXX). *Journal of Mental Deficiency Research,* 14(4):305-311, 1970.

A 20-month-old girl presented a rare chromosomal abnormality, quintuple-X (49,XXXXX) syndrome. The child exhibited developmental and mental retardation, abnormal facies, anomalies of limbs, congenital heart disease, and fingerprints with a total ridge count of only 33. Cause of this syndrome was probably nondysjunction in 2 consecutive meiotic divisions. This is the third reported case of quintuple X; main clinical signs in all cases were similar. (24 refs.) - *J. K. Wyatt.*

Psychoneurological Institute
Pruszkow, Poland

2067 **JANCAR, J.** Retinitis pigmentosa with mental retardation, deafness, and XX/XO sex chromosomes. *Journal of Mental Deficiency Research,* 14(4):269-273, 1970.

An SMR female (CA 73 yrs; IQ 33) presented retinitis pigmentosa in association with deafness, psychotic episodes, and dyskinesia. Chromosome analysis indicated XX/XO mosaicism; total ridge count on the finger tips was 161. Two brothers had defective sight and hearing, and one was also mute. The presence of dyskinesia, XX/XO mosaicism, and increased total ridge count may indicate that the condition is a sex-linked genetic expression in the female due to the absence of an X-sex chromosome in a proportion of cells. However, since mosaicism has occurred later in life in normal populations, it may not have been present at birth and may be incidental. (24 refs.) - *J. K. Wyatt.*

Stoke Park Hospital Group
Bristol, BS16IQU, England

2068 **REDDI, O. S.** Genetic effects of ^{131}I in mice. *Nature,* 227(5261):961-962, 1970. (Letter)

Mutagenic effects are reported for ^{131}I in mice. Males given the radioiodine intraperitoneally were mated with untreated females. Increased intrauterine deaths were observed during the first and fourth weeks. This suggests the possibility that ^{131}I may cause chromosomal breaks in spermatozoa and spermatocytes. (6 refs.) - *E. Kravitz.*

Osmania University
Hyderabad-7 (A.P.), India

2069 **SHAPIRO, LAWRENCE R.** Trisomy 18. *Lancet,* 2(7681):1035, 1970. (Letter)

Two cases of trisomy 18 are reported; they were observed between February and July of 1970. No other cases of this nature had been seen at this laboratory since its establishment in 1968. Confirmation of the diagnosis was by chromosome analysis. The incidence of trisomy 18 at this institution was calculated at 1:774 live births. (1 ref.) - *E. Kravitz.*

Mental Retardation Research Unit
Thiells, New York 10984

2070 **ARAKAKI, DAVID T.; & WAXMAN, SORRELL H.** Cytogenetics of spontaneous abortion: The chromosomes of decidua. *American Journal of Obstetrics and Gynecology,* 107(8):1199-1204, 1970.

Cytogenetic determinations made on cultured endometrial explants and fetal material from 68 spontaneous abortions revealed abnormal chromosomal endometrial karyotypes. Endometrial chromosomes were normal in the case of 22 chromosomally abnormal fetal membranes; in 4 cases both fetal membranes and endometria were chromosomally abnormal. The possibility that chromosomally abnormal endometrial cells increase with age is suggested. (16 refs.) - *E. Kravitz.*

Kauikeolani Children's Hospital
Honolulu, Hawaii 96813

2071 **EMBERGER, J.-M.; REY, J.; RIEU, D.; DOSSA, D.; BONNET, H.; & JEAN, R.** Trisomie du groupe C (47,XX,C+)[Group C trisomy (47,XX,C+)]. *Archives Francaises de Pediatrie,* 27(10):1081-1088, 1970.

Trisomy C was observed in a 5-year-old girl with significant psychomotor retardation, a dysmorphic syndrome affecting the face, trunk, and digits, dermatoglyphic anomalies, and radiological bone abnormalities, particularly in the spine. Chromosomal examination showed the presence of 47 chromosomes in all cells with an additional element in group C (6 to 12). Evidence of mosaicism was not manifested in any of 3 cultures. The Barr body was investigated in the buccal mucosa cells, and 40% of cells were found to possess a chromatin body. The fact that in 12 sufficiently marked mitoses a single chromosome of the C group and the X chromosome had a late replication confirmed that the excess chromosome was not an X chromosome. In a comparison of the clinical symptoms of the S with those in 13 cases reported previously, MR, facial dysmorphia, and congenital malformations were present in all. The most predominant features of trisomy C were found to include the absence of height and weight retardation and the presence of osseous abnormalities, speech limitations, minor genital anomalies, and muscular contractures. (13 refs.) - *B. J. Grylack.*

Centre de Transfusion sanguine
34-Montpellier, France

2072 **EMBERGER, J.-M.; REY, J.; RIEU, D.; DOSSA, D.; BONNET, H.; & JEAN, R.** Monosomie 21 avec mosaique 45,XX,21-/46,XX,21pi (Monosomy 21 with mosaic 45,XX,21-/46,XX,21pi). *Archives Francaises de Pediatrie,* 27(10):1069-1079, 1970.

Autoradiographically confirmed monosomy 21 with a mosaic of 45,XX,21-/46,XX,21pi in equal quantities was found in a girl aged 3½ years. Her birth weight was 1,940 g. No pathological symptoms were manifested in her parents. The S showed marked growth and psychomotor retardation, facial dysmorphia, dermatoglyphic defects, and radiological bone anomalies, primarily in the pelvic area. No deviation in platelets, leukocyte alkaline phosphatases, or in immunoglobulin (Ig) was revealed. The karyotype of the parents and 2 brothers was normal. Autoradiographic examination of the S disclosed the absence of a G chromosome with late replication. The triple mosaic can be explained by faulty division of the centromere with a loss of long acentric arms at the time of initial zygotic mitosis, leading to monosomy G and isochromy of the G short arms in equal amounts. The interpretation of an antimongoloidism and a monosomy 21 was favored, although antimongoloid obliquity of the palpebral fissures and biological anomalies were absent. (19 refs.) - *B. J. Grylack.*

Centre de Transfusion sanguine
34-Montpellier, France

2073 **EMBERGER, J.-M.; REY, J.; RIEU, D.; DOSSA, D.; BONNET, H.; & JEAN, R.** Interet de l'etude autoradiographique dans le diagnostic des anomalies chromosomiques rares: Un cas de trisomie 6-12(47,XX,C+); un cas de monosomie G (probablement 21) avec mosaique, 45,XX,G-/46,XX,Gpi/46,XX,Gqi (Significance of autoradiographic study in the diagnosis of rare chromosomal anomalies: A case of trisomy 6-12(47,XX,C+); a case of monosomy G (probably 21) with mosaicism, 45,XX,G-/46,XX,Gpi/46,XX, Gqi). *Archives Francaises de Pediatrie,* 27(8):874-875, 1970.

A 4-year-old girl presented with psychomotor retardation, normal height, facial dysmorphic syndrome, spinal bone anomalies as well as coxa vara, and a limitation of the extension of the fifth digits. Dermatoglyphics were abnormal. The karyotype revealed 47 chromosomes (47,XX,C+) without mosaicism, and the supplementary chromosome appeared to be one of the smallest of the group, perhaps a number 21. Autoradiographic examination disclosed a single X chromosome with late replication and only 1 Barr body in the cells of the buccal mucosa. A girl aged 3½ years manifested significant height and weight retardation and psychomotor deficiency with dysmorphia, as evidenced by microcephaly, hypotelorism, blepharochalasis, large and low-set ears, generalized contractures, and repeated vomiting without pyloric stenosis. The blood karyotype showed a mosaicism with 45,XX,G- and 46,XX,Gpi in equal proportions. Autoradiographic examination disclosed the lack of a G chromosome with late replication, including a number 21. Despite the absence of antimongoloid obliquity of the palpebral fissures and of biological anomalies, the other clinical and autoradiographic data favored a monosomy 21. (No refs.) - *B. J. Grylack.*

No address

2074 **SZCZEPSKI, O.; WALCZAK, M.; MACIEJEWSKI, J.; BITTNER, K.; CZEKALSKI, S.; & WALIGORA, A.** The behaviour of some indices of calcium-phosphate metabolism in Turner's syndrome. *Acta Paediatrica Scandinavica,* 60(1):73-77, 1971.

Clinical, radiological, hormonal, and cytogenic studies were performed on 6 patients with Turner's syndrome to evaluate some indices of calcium-phosphate metabolism and renal regulation of acid-base balance. Calcium and phosphate excretion were determined in feces collected for 3 consecutive days; calcium, phosphate, and creatinine concentrations in blood and urine were determined during loading tests; and the ability of the kidney to maintain acid-base balance was evaluated by the ammonium chloride test. Abnormalities revealed include a diminished end-organ sensitivity to parathormone, especially as expressed in terms of alterations of phosphate in renal tubules, which may affect growth and skeletal formation processes. There was an increased calcium excretion in the oldest subjects studied, which may be among the causes preventing pubertal growth acceleration. Some impairment of the efficient work of nephron was indicated by decreased hydrogen ion excretion before and after ammonium chloride loading. Ammonium ion excretion was also low in all but one subject. The function of the parathyroid glands appeared to be normal, but the reactivity of peripheral organs to this hormone may be involved in deformations of the osseous system and growth deficiencies. (15 refs.) - *N. Jarvis.*

IInd Clinic of Disease of Children
Medical Academy, Poznan, Poland

2075 **ALEXIOU, D.; CHRYSOSTOMIDOU, O.; VLACHOS, I.; & DELIGEORGIS, D.** Trisomy 18 with ovarian dysgenesis. *Acta Paediatrica Scandinavica,* 60(1):93-97, 1971.

A case of trisomy 18 with ovarian dysgenesis and additional pathologic findings is presented. The female infant was the child of an epileptic mother treated during pregnancy with diphenylhydantoin sodium and phenobarbitone. Multiple congenital abnormalities included odd-shaped head with prominent occiput and narrow forehead, broad and flat-bridged nose, low set misshapen ears, low hair line, small eyes, small mouth with hypoplastic mandible and high arched palate, upper extremities held at chest level with permanently clenched fingers and overlapping of index over middle and small over ring finger, hypoplastic thenar and hypothenar, small narrow pelvis, and short webbed toes. Chromosome studies revealed no abnormalities of the sex chromosomes; ovarian dysgenesis therefore seems to be another rare manifestation of trisomy 18. (13 refs.) - *N. Jarvis.*

State and University "Alexandra"
Maternity Hospital
Athens, Greece

2076 **RIDLER, M.A.C.; GARROD, O.; & BERG, J. M.** A case of Prader-Willi syndrome in a girl with a small extra chromosome. *Acta Paediatrica Scandinavica,* 60(2):222-226, 1971.

The appearance of the Prader-Willi syndrome, characterized by MR, short stature, obesity, hypogonadism, and muscular hypotonia, is described in a patient with a small extra chromosome. The extra chromosome was slightly smaller and more metacentric than the chromosomes of the G group and in some cells was accompanied by satellites, whose presence suggests an origin from the D or G group of chromosomes. Seven instances of chromosomal abnormality have been reported among 58 cases karyotyped, but further data must be accumulated before the significance of this and other findings can be evaluated. (12 refs.) - *N. Jarvis.*

Harperbury Hospital
St. Albans, Hertfordshire, England

2077 Is there a 'typical' XYY personality? *Medical World News,* 12(3):40, 1971.

Recent discussions of the relationship between genetics and human behavior have cast doubt on assumptions concerning the association of antisocial behavior with an extra Y chromosome in males. A review of reported cases of XYY males seems to imply that tallness is the only common characteristic. Despite numerous surveys, no one is certain about the prevalence of the double Y chromosome in the general population. A crude

estimate claims the presence of 1 to 2 XYY individuals among every 1,000 males. This prediction has led some people to conclude that there are thousands of dangerous individuals at large. Much more knowledge must be gained, however, before preventive legal measures are taken with regard to this genetic class. (No refs.) - *B. J. Grylack.*

2078 **CH'ENG, LESLIE Y.; & WALZ, DONALD V.** "Cri du Chat" syndrome. Report of a case. *Ohio State Medical Journal,* 67(3):237-240, 1971.

An 8-year-old white girl admitted to an MR center presented with profound MR, microcephaly, hypertelorism, low-set ears, and growth retardation. Chromosome analysis of peripheral lymphocytes showed a karyotype of 46,XX-5p, with a deletion of short arms of 1 chromosome of B group, probably number 5. On the basis of these findings, plus the history of a cat-like cry, a diagnosis of "Cri du Chat" syndrome was made. The mother had taken several medications prior to and during her pregnancy with this child, but none is known to be related to chromosomal aberrations. Karyotypes of both parents of the child were normal. The child will continue to live at home with her family, a situation which will maximize her potential development. (20 refs.) - *B. J. Grylack.*

Broadview Center for the Mentally Retarded
Broadview Heights, New York

2079 **DISHOTSKY, NORMAN I.; LOUGHMAN, WILLIAM D.; MOGAR, ROBERT E.; & LIPSCOMB, WENDELL R.** LSD and genetic damage. *Science,* 172(3982):431-440, 1971.

Of 9 studies *in vitro* of the correlation between lysergic acid diethylamide (LSD) and genetic damage, 6 have indicated the existence of some induced chromosomal breakage, while 3 have failed to confirm this finding. In 21 chromosomal studies *in vivo,* 126 Ss were treated with pure LSD, and 184 were exposed to illicit, "alleged" LSD. A maximum of only 14.29% of Ss in the pure LSD group showed higher frequency of chromosomal aberration than the controls. In contrast, a maximum of 48.91% of Ss in the illicit LSD group manifested an increase in the frequency of aberrations. On the basis of these and related observations, it appears that pure LSD ingested in moderate doses does not damage chromosomes *in vivo,* does not cause detectable genetic damage, and is neither a teratogen nor carcinogen in the human. Except during pregnancy, there is no present contraindication to the continued controlled experimental use of pure LSD. (101 refs.) - *B. J. Grylack.*

Mendocino State Hospital
Talmage, California

2080 **RUDIGER, H. W.; PASSARGE, E.; HIRTH, L.; GOEDDE, H. W.; BLUME, K. G.; LOHR, G. W.; BENOHR, H. C.; & WALLER, H. D.** Triosephosphate isomerase gene not localized on the short arm of chromosome 5 in man. *Nature,* 228(5278):1320-1321, 1970. (Letter)

Five persons with partial deletion of the short arm of chromosome 5 and normal triosephosphate isomerase (TPI) are described; this suggests that localization of the gene for TPI on the short arm of chromosome 5 is not likely. All 5 patients (ages 1 to 9 years) exhibited the cat cry syndrome, including psychomotor retardation. Chromosome studies on lymphocytes showed significant variation in the extent of the short-arm deletion. (5 refs.) - *E. Kravitz.*

Universitat Hamburg
2000 Hamburg 20, Germany

2081 **FIALKOW, PHILIP J.; THULINE, HORACE C.; HECHT, FREDERICK; & BRYANT, JEAN.** Familial predisposition to thyroid disease in Down's syndrome: Controlled immunoclinical studies. *American Journal of Human Genetics,* 23(1):67-86, 1971.

Families of children with Down's syndrome, both at home and in institutions, were evaluated for the frequency of thyroid disease. Clinical thyroid disease was found in 30 (17%) of 177 mothers of patients with Down's syndrome and in 11 (6%) of 177 control mothers (children with other chronic illnesses or MR). Fourteen of the 30 mothers had the disease documented before the

birth of the affected children. Seven of 70 sisters of subjects had thyroid disease, while none of the 66 control sisters did. Ten of 159 maternal aunts of subjects also had thyroid disease. Significant increases in the frequency of thyroid antibodies were found in Ss with Down's syndrome and in their mothers. The relationship between thyroid autoimmunity and Down's syndrome appears established; however, the reason for this has yet to be determined. (28 refs.) - *E. L. Rowan.*

University of Washington
Seattle, Washington 98105

2082 ELLER, ELEANOR; FRANKENBURG, WILLIAM; PUCK, MARY; & ROBINSON, ARTHUR. Prognosis in newborn infants with X-chromosomal abnormalities. *Pediatrics,* 47(4):681-688, 1971.

Routine screening of 21,214 consecutive newborns for chromatin constitution revealed 32 with X chromosome abnormalities. These infants have been followed for 1½ to 5 years with periodic physical and developmental examinations, psychological testing, and social evaluations in order to determine the prognosis for these anomalies. Nine of the infants showed a 47,XXX abnormality, 8 47,XXY, 3 45,X, and the rest were mosaics. Three died in infancy. The triple X group commonly showed clinodactyly, epicanthal folds, and wide-set eyes, and the group with Turner's syndrome had classical signs, but otherwise the children were phenotypically normal. Only 2 of the children are MR, but 2 others show difficulty which may be cultural in origin. Many have a tendency toward emotional difficulty, such as shyness or depression. Followup chromosomal analysis in mosaicism shows a tendency for aneuploid lines to decrease. This may explain the phenotypic appearance of some older patients with normal chromosomes. (31 refs.) - *E. L. Rowan.*

University of Colorado Medical Center
Denver, Colorado 80220

2083 MATTHEW, EARL B.; DIETZMAN, DALE E.; MADDEN, DAVID L.; SEVER, JOHN L.; AL-AISH, MATTI S.; & DODSON, W. EDWIN. Possible absence of Australia antigen in G/G translocation-type Down's syndrome. *Lancet,* 1(7701):702, 1971. (Letter)

The incidence of Australia antigen was determined in the various karyotypes of Down's syndrome. Six of 7 subjects with trisomy 21 were found to carry the antigen; neither of 2 patients with G/G translocation carried it. Two of 14 normal controls had the antigen. The relationship is suggestive, but the number of tested patients with translocations is as yet too small. Pooled resources are suggested in order to determine if this represents a meaningful genetic relationship. (2 refs.) - *E. L. Rowan.*

National Institutes of Health
Bethesda, Maryland 20014

2084 JOHNSON, CHARLES F. The thumb in trisomy-18. *Journal of the American Medical Association,* 215(3):489, 1971. (Letter)

A letter to the editor regarding an abnormal hand which simulated trisomy-18 failed to emphasize that the patient's thumb appeared normal. In trisomy-18 the thumb is high-set and short with an ulnar clinodactyly which may cause it to overlap the second digit. The hypoplasia of the thenar eminence is marked and has been shown to be due to aplasia of intrinsic thenar musculature. This high position of the thumb persists even when the overlapping finger anomalies disappear in time. (2 refs.) - *J. C. Moody.*

No address

2085 STENCHEVER, MORTON A.; & JARVIS, JANE A. Diphenylhydantoin: Effect on the chromosomes of human leukocytes. *American Journal of Obstetrics and Gynecology,* 109(6):961-962, 1971.

The effects of diphenylhydantoin (Dilantin), an anticonvulsant drug, on the chromosomes of human leukocytes *in vitro* were investigated in 2 cultures each of 100.0, 10.0, 1.0, and 0.1 μg diphenylhydantoin per ml of culture fluid plus 2 control cultures of heparinized venous blood drawn from 2 individuals in good health with no recent exposures to agents known to break chromosomes. Slides were prepared, coded, and

scored for chromatid and isochromatid gaps and breaks and for abnormal forms. There were no significant differences between the data for the various concentrations and the controls. Although diphenylhydantoin does not appear to cause obvious chromosome damage, it should be remembered that the drug does seem to be teratogenic in mice and that it may, under certain conditions, have a similar effect in human beings. (3 refs.) - *J. C. Moody.*

50 N. Medical Dr.
Salt Lake City, Utah 84112

2086 FEINGOLD, MURRAY; GHERADI, GHERARDO J.; & SIMONS, CHRISTINE. Familial neuroblastoma and trisomy 13. *American Journal of Diseases of Children,* 121(5):451, 1971. (Letter)

A case illustrates a hitherto unknown association of familial neuroblastoma and trisomy 13. The patient was a 2½-year-old white girl who had a large neuroblastoma of the left adrenal gland. After 3 months of treatment with various drugs, including procarbazine hydrochloride, the patient died. A year later her mother gave birth to a child with multiple anomalies, who died at age 2 days. Examination of the infant revealed all the characteristics of trisomy 13, confirmed by chromosomal analysis, as well as numerous neuroblastoma. Since the mother had tasted the procarbazine hydrochloride medication 2 to 3 months before conception of the second child, the possibility of a teratogenic effect from the drug exists. Some correlation may obtain between familial neuroblastoma and trisomy 13. (3 refs.) - *S. Glinsky.*

Boston Floating Hospital for
Infants and Children
Boston, Massachusetts 02111

2087 AL-AISH, MATTI S.; DODSON, EDWIN; & PLATO, CHRIS C. Down's syndrome with XYY:48,XYY,G+. *American Journal of Diseases of Children,* 121(5):444-446, 1971.

A rare case of a patient with trisomy 21 and XYY is described. The patient was a 9-month-old boy, had a karyotype of 48,XYY,G+, and presented with the usual characteristics of Down's syndrome, except for the absence of epicanthal folds and a prominent occiput. This condition would be expected to occur at a frequency of 1:480,000 male births, based on the assumption that the 2 events occur independently. Analysis of blood groups, haptoglobins, and dermatoglyphics was unrevealing. No clear modifications of the mongoloid phenotype by the additional Y were apparent. (21 refs.) - *S. Glinsky.*

National Institute of Child Health
and Human Development
Bethesda, Maryland 20014

2088 EBBIN, ALLAN J.; & WILSON, MIRIAM G. Trisomy 18. *Lancet,* 1(7689):77-78, 1971. (Letter)

Fluctuations in the incidence of rare conditions are substantially affected by the investigator's choice of the time-period for study. A recent study by other researchers reported apparent trisomy 18 increases, but these seem due to a biased time-period selection procedure. The present study surveyed birth defects in 8 hospitals between July, 1969, and November, 1970, and determined a trisomy 18 incidence of 1 in 6,000 live births. Restricting consideration to the one month of January, 1970, yields an incidence of 1 in 900 live births. The results indicate the necessity for continuous surveillance over a long period to assess incidence increases accurately. (2 refs.) - *S. Glinsky.*

Los Angeles County-U.S.C. Medical Center
Los Angeles, California 90033

2089 MANTLE, D. J.; PYE, CAROLE; HARDISTY, R. M.; & VESSEY, M. P. Plasma factor-VIII concentrations in XXX women. *Lancet,* 1(7689):58-59, 1971.

Factor-VIII assays of plasma samples from XXX women provided no evidence for inactivation of the X-borne factor-VIII locus. Assays were performed on 2 to 4 samples from each of 12 XXX women and 12 age-matched normal controls. In each case the mean value was higher for the XXX S than for her matched control, and the overall mean difference between the log concentrations was highly significant. Since most other

influences on plasma factor-VIII concentration were eliminated by the investigation's design, blood group differences, anxiety at venepuncture, and inherent assay technique errors remain as possible sources of variability. However, the possession of an extra X chromosome seems likely as responsible for some of the difference observed. The way in which the extra X chromosome exerts its effect on the factor-VIII concentration remains unexplained. 25 refs. - *S. Glinsky.*

Health Department, Municipal Offices
Rotherham, Yorkshire, England

2090 **TIMSON, J.; HARRIS, R.; GADD, R. L.; FERGUSON-SMITH, M. E.; & FERGUSON-SMITH, M. A.** Down's syndrome due to maternal mosaicism and the value of antenatal diagnosis. *Lancet,* 1(7698):549-550, 1971. (Letter)

Inheritance of Down's syndrome from a mother exhibiting mosaicism is reported, bringing the number of known cases of such mothers who have borne children with trisomy 21 to 8. A pregnant 36-year-old mother, who had already given birth to a male child with Down's syndrome at age 26, revealed no abnormalities upon examination except a 10% blood karyotype of 47,XX,21+. Analysis of cultured amniotic cells determined them to be 46,XX. The pregnancy went to term and the baby was clinically and cytogenetically normal. Adding this case to the 7 reported by Aarskog, 8 mothers with mosaicism have given birth to 11 children with Down's syndrome and 2 normal children. After removal of the propositi, the empirical recurrence risk is 3 in 5. Where this kind of mosaicism has been detected in a pregnant woman it is important to attempt antenatal diagnosis. (6 refs.) - *S. Glinsky.*

St. Mary's Hospital
Manchester M13 OJH, England

2091 **KUCERA, JIRI.** Infectious hepatitis and Down's syndrome. *Lancet,* 1(7698):549, 1971. (Letter)

Objections to the author's study linking infectious hepatitis and Down's syndrome are based on supposition, not facts. The likelihood that false-positive, false-negative diagnoses of infectious hepatitis and memory biases would compromise the results is minimal, since in Czechoslovakia suspected cases of infectious hepatitis require hospital admittance, evaluation by hygiene departments, and strict dieting and repeated tests. In those countries where no association was found, the rate of notification of infectious hepatitis cases is very small or nil. Population density may also considerably influence association. Suggestions made regarding improved investigation design features would yield more consistent results. (5 refs.) - *S. Glinsky.*

Ustav pro peci o matku a dite
Praha 4-Podoli, Czechoslovakia

2092 **SERGOVICH, F.; UILENBERG, CATHARINA; & POZSONYI, J.** The 49,XXXXX chromosome constitution: Similarities to the 49,XXXXY condition. *Journal of Pediatrics,* 78(2):285-290, 1971.

A 16-year-old girl is described who is the fourth case of pentaploidy-X chromosome constitution noted in medical literature. Examination and cytogenetic analysis of leukocyte and skin cultures revealed 49 chromosomes, up to 4 sex chromatin masses in interphase nuclei, and a penta-X chromosome constitution. The patient presented a distinctive facies which was remarkably similar to that of a girl with a penta-X/tetra-X mosaic constitution. Also both girls exhibited a large number of skeletal malformations identical to those found in males possessing tetra-XY sex chromosomes. It is suggested that this large overdose of chromosomes caused the carriers in both males and females to share common pathways in the development of the skeletal systems. (5 refs.) - *S. Glinsky.*

Children's Psychiatric Research Institute
London, Ontario, Canada

2093 **COFFIN, GRANGE S.; & WILSON, MIRIAM G.** Wolf-Hirschhorn syndrome. *American Journal of Diseases of Children,* 121(3):265, 1971. (Letter)

Chromosome reanalysis indicates that 2 of 3 patients reported earlier as having normal chromosomes have in fact partial deletion of the

short arm of a Number 4 chromosome (4p-) characteristic of the Wolf-Hirschhorn syndrome. Chromosome analyses of the third patient and his parents remain normal. Since all 3 patients present with the other characteristic manifestations of the Wolf-Hirschhorn syndrome, the chromosome-normal child may have submicroscopic deletion of chromosome No. 4, another cytogenetic mechanism, or a similar phenotype but from an unidentified cause. (3 refs.) - *S. Glinsky.*

No address

2094 KADOTANI, TETSUJI; OHAMA, KOSO; SOFUNI, TOSHIO; & IKEDA, TAKAYOSHI. An XY human female with ovaries. *American Journal of Obstetrics and Gynecology*, 110(1):98-102, 1971.

A case of a phenotypic female with afollicular ovarian dysgenesis and an XY chromosome constitution is described. The patient, a 31-year-old female, was hospitalized with primary amenorrhea and was studied cytogenetically. The patient was diagnosed as having afollicular ovarian dysgenesis, with the presence of a vagina, a uterus, bilateral Fallopian tubes, and the absence of testes as well as the somatic signs of Turner's syndrome. Chromosome studies carried out on cultured leukocytes and fibroblasts from various organs indicated an apparently normal male chromosome constitution of 46,XY. Also the deoxyribonucleic acid-replicating pattern and the fluorescent-staining property observed in the cultured leukocytes were consistent with that of normal male cells. Intelligence testing showed a Wexler Adult Intelligence Scale score of 65 and a Tanaka-Binet score of 67.7. The XY chromosome constitution may derive from the testis-determining effect of the Y chromosome being either adversely affected by some environmental factor, or suppressed by an abnormal gene or genes. (16 refs.) - *S. Glinsky.*

Hiroshima University School of Medicine
Hiroshima, Japan

2095 HARRIS, JEROME S.; & ROBINSON, ARTHUR. X chromosome abnormalities and the obstetrician. *American Journal of Obstetrics and Gynecology*, 109(4):574-583, 1971.

X chromosome abnormalities frequently are associated with many important medical and psychosocial problems and can be detected before the associated problem is apparent. The incidence, significance, and cause of X chromosome abnormalities were determined through a longitudinal study of 21,214 unselected newborn infants of at least 28 weeks gestation. The Ss were screened for the presence of abnormalities by Barr body counts, nuclear sexing, and nuclear sex-phenotypic sex comparison. The screening methods utilized buccal smears for the first 1,475 cases and amniotic membrane for the remainder. The results indicated Barr body counts outside of the normal range, phenotypic sexual ambiguity, and a clear discrepancy between nuclear sex and phenotypic sex. X chromosome abnormalities did not seem related to socioeconomic status, maternal age, or type of obstetric care, but did appear related to the season of the year at time of conception and to an intercurrent rubella epidemic. The early detection of X chromosome abnormalities is of great value to the individual family, physician, and the community, and may most easily be effected by the routine nuclear sexing of all newborn infants. (45 refs.) - *S. Glinsky.*

Sinai Hospital of Baltimore
Baltimore, Maryland 21215

2096 STENCHEVER, MORTON A.; & JARVIS, JANE A. Cytogenetic studies in reproductive failure. *Obstetrics and Gynecology*, 37(1):83-89, 1971.

Individuals can possess karyotypic abnormalities compatible with phenotypic normality but not with successful reproduction. The Ss in this study were 28 couples and 3 individuals suffering from problems of reproductive failure. Of these, 3 were identified as having chromosome abnormalities. The 3 were a female carrier of a heterologous D/D translocation (13/15), a female with an XX/XXX mosaic pattern, and a male with a 47,XYY karyotype, the most common sex chromosome anomaly. Since individuals with such abnormalities might be expected to produce conceptuses with a greater number of chromosome anomalies than would the general population, perhaps leading to frequent abortions, stillbirths, congenital malformations, and MR, their location may have important preventive benefits. (32 refs.) - *S. Glinsky.*

University of Utah Medical Center
Salt Lake City, Utah 84112

2097 **TURNER, GILLIAN; TURNER, BRIAN; & COLLINS, EDITH.** Renpenning's syndrome: X-linked mental retardation. *Lancet*, 2(7668):365-366, 1970. (Letter)

Five families are reported with a history of X-linked mental retardation among 25 males. Of these, the 14 examined had no physical signs; there was moderate retardation (IQ 35-51) in most cases. Records of 148 additional males with moderate retardation were also studied; 17 of these males had no abnormality, but in the case of 11 out of 17 there was either an affected male sibling or a family history suggesting X-linkage. The lack of physical abnormality in a moderately retarded male suggests X-linked mental retardation (Renpenning's syndrome). (2 refs.) - *E. Kravitz.*

No address

2098 **DOXIADIS, S. A.; PANTELAKIS, S. N.; & VALAES, T. N.** Infectious hepatitis and Down's syndrome. *Lancet*, 2(7677):826, 1970. (Letter)

Explanatory comments are offered on a previous paper which suggested a possible correlation between Down's syndrome and infectious hepatitis, and a critical letter concerning this previous paper is answered. The small number of subjects involved made it desirable to confirm the preliminary results reported. A questionnaire had been used to develop the medical history of each subject; possible effects of maternal age and family size were considered. (3 refs.) - *E. Kravitz.*

Queen Anna-Maria Institute of Child Health
Athens 608, Greece

2099 **KRMPOTIC, EVA; ROSNICK, MICHAEL R.; & ZOLLAR, LOWELL M.** Genetic counseling: Secondary nondisjunction in partial trisomy 13. *Obstetrics and Gynecology*, 37(3):381-390, 1971.

A mother and daughter had identical minute extra chromosomes, a deleted number 13 chromosome. The child had gross abnormalities, failed to thrive, and died at the age of 80 days. Some phenotypic abnormalities were similar in the mother but in a milder form. She presented a slight antimongoloid slant of the eyes and hypertelorism, deformed right tragus with preauricular tags, and a draining preauricular sinus. Neither the father nor the 4 siblings had physical deformities or abnormal karyotypes. The extra chromosome in the affected child resulted from secondary nondisjunction during meiosis in the maternal germ cell. Genetically high-risk pregnancies should be identified and monitored. Cytogenetic diagnosis before birth can provide exact prenatal diagnosis. (29 refs.) - *J. K. Wyatt.*

Mount Sinai Hospital Medical Center
Chicago, Illinois 60608

2100 **BIRNBAUM, STANLEY J.** Prenatal diagnosis of mongolism by X-ray. *Obstetrics and Gynecology*, 37(3):394-395, 1971.

An abnormal fetal attitude which consists of hyperextension of the head with dorsiflexion of the cervicodorsal vertebrae observed in abdominal X-rays taken during the third trimester of pregnancy is highly suggestive of a fetus with Down's syndrome. Retrospective analysis of X-rays taken near term of 5 patients who later delivered mongoloid infants revealed this abnormal fetal attitude in all cases. These data were later used to predict the birth of a mongoloid infant to a mother who was admitted to hospital at term because of toxemia. (3 refs.) - *J. K. Wyatt.*

Brookdale Hospital Center
Brooklyn, New York 11212

2101 **HALBRECHT, I.; KLETZKY, O.; KOMLOS, L.; LOTKER, L.; & GERSHT, N.** Trisomy-D cyclops: A case report. *Obstetrics and Gynecology*, 37(3):391-393, 1971.

A female newborn exhibited cyclops associated with trisomy D (47,XXD+). Chromosomal analysis of the parents and a normal brother revealed normal karyotypes. Although most published cases of females record a high percentage of genital malformations, none were detected in this patient at autopsy. There appear to be several different malformations and karyotypes associated with cyclops. (13 refs.) - *J. K. Wyatt.*

Hasharon Hospital
Petah Tiqva, Israel

2102 LINDENBAUM, R. H.; & BUTLER, L. J. Child with multiple anomalies and a group B(4-5) long arm deletion (Bq-). *Archives of Disease in Childhood,* 46(245):99-101, 1971.

A male child who seemed to present a new clinical entity had multiple anomalies and a 46,XY,B5q- karyotype. The parents were normal, healthy, and unrelated and had no family history of MR or congenital abnormality. At birth, the child had an oddly shaped skull, pitting edema of hands and feet, and became limp and cyanosed during the first 8 hours of life. He had an abnormal skull with brachycephaly, a very large and sunken anterior fontanelle, a widely open metopic suture, prominent frontal bones, parietal bones contiguous across the sagittal suture, and a large posterior fontanelle. His eyes were deeply set and small, the mandible was small, and the ears were slightly low set. He had short arms with ulnar angulation at the wrists and grossly incurved little fingers. He had respiratory difficulties until the age of 4 weeks. The femora seemed short, and the right testicle was undescended. He remained hypotonic and unresponsive. At 9 months, just prior to death from an acute respiratory illness, he adopted a persistently supine position, showed complete head lag, and had never been observed to smile. Visual and auditory responses demonstrated little development since the neonatal period. Necropsy showed signs of chronic pulmonary infection, diffuse polymicrogyria in the brain, and agenesis of the corpus callosum. Brain weight was 510g. He had deficient cerebrospinal fluid and an enlarged right kidney. The mother has since given birth to a normal boy. (4 refs.) - *J. K. Wyatt.*

Westminster Hospital
London SW1, England

2103 MIR, G. H.; & CUMMING, GORDON R. Response to atropine in Down's syndrome. *Archives of Disease in Childhood,* 46(245):61-65, 1971.

Heart rate response to atropine was similar in MR patients with and without Down's syndrome. Doses of 30 to 60 mg/kg of atropine were administered to 7 male and 7 female children with Down's syndrome (CA range 2 to 10 yrs), 5 boys and 2 girls with chronic brain damage (CA range 3 to 10 yrs), 6 male and 5 female adults with Down's syndrome (CA range 20 to 29 yrs), and 5 men and 4 women with MR of unknown origin (CA range 22 to 32 yrs). After the initial 7 mg/kg of atropine, heart rate decreased below control values for both groups of children and then increased to a mean maximum rate of 153 beats/minute for control children at dosage of 35 mg/kg of atropine and to 143 beats/minute for mongoloid children. Mean maximum change in heart beat rate occurred at an atropine dosage of 30 mg/kg for control adults and at 25 mg/kg for mongoloid adults. Heart rates of Down's syndrome Ss were significantly lower ($p<.05$) than those of control Ss when propranolol was given in addition to atropine. Down's syndrome Ss exhibited more rapid mydriasis in response to ophthalmic atropine. (10 refs.) - *J. K. Wyatt.*

Children's Hospital
Winnipeg, Canada

2104 CATTI, A.; & SCHMID, W. A pericentric inversion, 5p-q+, and additional complex rearrangements in a case of cri-du-chat syndrome. *Cytogenetics,* 10(1):50-60, 1971.

The karyotype of a girl with the characteristic clinical features of the cri-du-chat syndrome shows 3 abnormal chromosomes: a 5p-q+, a ring C, and a 15q+. The cri-du-chat phenotype appears due to a submicroscopical lesion on the distal segment of the short arm of chromosome 5, as a pericentric inversion seems to have altered a critical section. The additional structural rearrangements apparently have no effect on phenotype. The mother of this subject shows acentric fragments and ring chromosomes in 5.3% of 225 analyzed cells. (8 refs.) - *E. L. Rowan.*

Hopital Cantonal Universitaire
1011 Lausanne, Switzerland

2105 TETTENBORN, U.; GROPP, A.; MURKEN, J.-D.; TINNEFELD, W.; FUHRMANN, W.; & SCHWINGER, E. Meiosis and testicular histology in XYY males. *Lancet,* 2(7666):267-268, 1970.

Lymphocyte cultures from 4 XYY males revealed a 47,XYY karyotype without any evidence of somatic mosaicism. The 4 male patients were 16,

18, 23, and 35 years of age and were characterized respectively by no psychiatric abnormality, educational subnormality, and institutional commitment for sexual assault and violence. Histological examination in all 4 cases revealed different degrees of tubular atrophy with impairment of spermatogenesis before or at least beyond the secondary-spermatocyte level. Case 1 had no spermatocytes, while the majority of the 67 metaphases from the other 3 cases displayed a 47,XYY karyotype. Cases 2 and 4 provided primary spermatocytes of good figure quality, permitting unequivocal identification of bivalents. An alternative explanation of the elimination of the Y chromosome may be the random loss of one of the Y chromosomes during the spermatogonial divisions. A proliferative advantage of the 46,XYY germ-cells may play an additional important role. (15 refs.) - *S. Glinsky.*

Universitaet Bonn/Rhein
Bonn, West Germany

2106 SIVA SANKAR, D. V. Chromosome breakage in infantile autism. *Developmental Medicine and Child Neurology,* 12(5):572-575, 1970.

Cultured leukocytes from autistic children show a considerably larger number of chromosome breakages than do those from nonautistic children. The Ss were 67 children, of whom 18 were diagnosed as nonautistic schizophrenic, 31 as autistic schizophrenic, 9 as psychotic, and 9 as afflicted with behavioral disorders. Chromosome analysis revealed no significant aneuploidy or mosaicism in any of the groups, but chromosome breakages in autistic children were considerably greater than breakages in nonautistic children. This finding, taken in conjunction with the fact that the autistic child differs from the nonautistic child in many biochemical parameters, may indicate that infantile autism is an extensive pathological disorder. (11 refs.) - *S. Glinsky.*

Queens Child Psychiatric Hospital
Creedmore State Hospital
New York, New York 11427

2107 WILLIAMS, JERRY D.; & SUMMIT, ROBERT L. A review of clinical symptoms associated with aberrations of the autosomes. *Journal of the Tennessee Medical Association,* 64(4):310-317, 1971.

Syndromes resulting from autosomal anomalies are characterized by low birth weight, failure to thrive, MR, and congenital anomalies related to multiple organ systems. Autosomal anomalies occur because of numerical or structural errors. The most common numerical error is trisomy. This includes trisomy 21 or Down's syndrome, 18-trisomy or Edward's syndrome, and 13-trisomy or Patau's syndrome. Among the structural errors is the cri du chat syndrome. Clinical descriptions of these anomalies are included. (No refs.) - *J. K. Wyatt.*

University of Tennessee Medical Unit
Memphis, Tennessee

2108 REED, TERRY E.; CHRISTIAN, JOE C.; & NANCE, WALTER E. Diagnosis of mongolism with a dermatoglyphic nomogram. *Southern Medical Journal,* 64(1):70-72, 1971.

Analyses of dermatoglyphic findings on 250 karyotypically proven trisomic mongoloids and 332 individuals with normal karyotypes were used to develop a systematic method for diagnosis of mongolism. Frequencies of dermatoglyphic pattern types were identified in 32 pattern areas, and likelihood ratios were calculated for each pattern type in each pattern area. The sign and magnitude of the score yielded by the logarithm of these ratios were used to diagnose the presence of Down's syndrome or a normal karyotype. (4 refs.) - *J. K. Wyatt.*

Indiana University School of Medicine
Indianapolis, Indiana 46202

2109 KERSEY, J. H.; YUNIS, J. J.; LEE, J.C.; & BENDEL, R. P. Antenatal detection of partial C (6-12) trisomy. *Lancet,* 1(7701):702-703, 1971.

A karyogram of cells cultured from amniotic fluid withdrawn at 18 weeks gestation during the third pregnancy of a woman whose karyotype was normal but whose husband had a short C and a long D chromosome, representing a C/D

balanced translocation, demonstrated 46 chromosomes with an XX sex complement and an abnormally long arm of a D (13-15) chromosome representing a partial C (6-12) trisomy. Subsequent instillation of hypertonic saline delivered a stillborn female fetus which, at necropsy, was found to have retarded growth, agyria, low-set ears, macroglossia, micrognathia, high-arched palate, webbing of the neck, hypoplastic left kidney, hypoplastic adrenals, muscular defect of the left side of the diaphragm, bilateral simian crease, campodactyly along with interphalangeal crease between the fourth and fifth fingers, severe bilateral talipes equinovarus, and webbing between the second and third toes of the left foot. The necropsy findings thus confirm the antenatal diagnosis of partial C (6-12) trisomy, representing the same syndrome as that in the patient's living 4-year-old, multiply handicapped, SMR son from her second pregnancy. The first pregnancy had ended in spontaneous abortion at 4 months. (8 refs.) - *J. C. Moody.*

University of Minnesota
Minneapolis, Minnesota 55455

2110 BUTLER, L. J.; & REISS, H. E. Antenatal detection of chromosome abnormalities. *Journal of Obstetrics and Gynaecology of the British Commonwealth*, 77(10):902-907, 1970.

An abdominal amniocentesis procedure, feasible during the fourteenth to sixteenth weeks of pregnancy, to obtain cells withdrawn from the 15-20 ml amniotic fluid for subsequent culture and karyotyping is described. The culture techniques detailed are designed to alleviate difficulties arising from the tendency of viable cells to attach themselves rapidly to glass surfaces. Culture time necessary to produce chromosome preparation averaged 4 weeks with a range from 2 to 8 weeks. The relative success rate of culturing cells and producing karyotypes was 88%. Because even in a rich amniotic fluid cell culture, the total cells usually represent only one-tenth of the population of a comparable fibroblast culture, and because there is poor survival of cells of many cultures after only 2 or 3 passages, it is unlikely that routine screening for a large number of enzyme defects will be feasible. With a 4-6 weeks' wait for results, problems of arousing undue anxiety by testing otherwise contented pregnant women when the chance of chromosome abnormality is still fairly small should also be considered. Amniotic fluid cell culture investigations are therefore only suitable for screening the high risk pregnancy. On economic grounds, however, the cost of carrying out 100 amniocentesis tests on women over 40 to identify 1 mongol is but one-fifth of the cost of maintaining a Down's syndrome patient in an institution for a lifetime. (24 refs.) - *J. C. Moody.*

Queen Elizabeth Hospital for Children
London E.2, England

MEDICAL ASPECTS — Miscellany

2111 CRITCHLEY, E.M.R. Naturally occurring dyskinesias. *Lancet*, 2(7682):1082-1083, 1970. (Letter)

Naturally occurring movement disorders (dyskinesias) may be concomitants of organic disease or degeneration, or they may occur in isolation. The latter group has often been considered to be part of nonspecific MR or psychiatric syndromes, but these should be reevaluated in light of recent discoveries concerning the effects of dopamine and other central nervous system transmitters. (4 refs.) - *E. L. Rowan.*

No address

2112 OLNEY, JOHN W.; & HO, OI-LAN. Brain damage in infant mice following oral intake of glutamate, aspartate or cysteine. *Nature*, 227(5258):609-611, 1970. (Letter)

Toxic reactions were produced in infant mice fed glutamate or aspartate at low levels (0.5 gm/kg glutamate; 1 gm/kg aspartate). An additive brain-damaging effect was observed after feeding both of these amino acids (each at 0.5 gm/kg). Both of these substances are neuroexcitatory, and they produced hypothalamic damage (e.g., to the arcuate nucleus). Aspartate also produced retinal injury. Cysteine (3 gm/kg) p.o. induced both

hypothalamic and retinal damage. No toxic reactions resulted from the ingestion of any other amino acid. The studies suggest serious doubt as to the advisability of introducing monosodium glutamate into the diet of the human infant. (12 refs.) - *E. Kravitz.*

Washington University School of Medicine
St. Louis, Missouri 63110

2113 **SMITH, G. F.; SCHINDELER, JOAN; ELBUALY, S.; & SHEAR, CAROL.** Limb abnormalities and mental retardation. *Journal of Mental Deficiency Research,* 14(4):319-341, 1970.

Although MRs frequently present abnormalities of limbs, especially upper limbs, the association between abnormal limb development and brain is generally not explainable and is poorly understood. Of 18 MRs with limb abnormalities, 10 were classified in known syndromes: 3 cases of Laurence-Moon-Biedl and 1 case each of Apert, Marfan, de Lange, Rubinstein-Taybi, Holt-Oran, partial trisomy, and unilateral limb deformities and psoriasiform dermatosis. None of these families had another affected sibling or parent. In the other 8 cases, 2 patients had MR siblings who did not have associated limb abnormalities, and 2 patients had relatives with limb abnormalities who were not MR. Even in syndromes where limb abnormalities regularly occur, expression of the limb deformity varies markedly. (15 refs.) - *J. K. Wyatt.*

Loyola University
Maywood, Illinois 60153

2114 **SILVER, LARRY B.** A proposed view on the etiology of the neurological learning disability syndrome. *Journal of Learning Disabilities,* 4(3):123-133, 1971.

Research data on 1 group of children with a neurological learning disability syndrome suggest that this syndrome may be the result of a neurohumoral deficiency: a primary dysfunction of the ascending reticular activating system and a secondary dysfunction of the limbic system. These children have difficulty in the development of satisfactory relationships with their environment, and in receiving, processing, storing, and expressing information. They also develop emotional problems. The clinical syndrome presented is improved by drugs which increase available central nervous system norepinephrine. Clinical experience in treating children with medication suggests that when 2 drugs, each capable of increasing the amount of available norepinephrine in the brain, were given concurrently, an "overreaction" occurred. Other research suggests the presence of a genetic factor where a type of nervous system in which the pathology may be due to neurohumoral imbalance is inherited. (46 refs.) - *J. K. Wyatt.*

The Psychiatric Institute
Washington, D. C. 20037

2115 **PICKERSGILL, MARY J.; & PANK, PENELOPE.** Relation of age and mongolism to lateral preferences in severely subnormal subjects. *Nature,* 228(5278): 1342-1344, 1970. (Letter)

Effects of age and mongolism on handedness (lateral preference) were studied in 32 SMR hospitalized patients who were able to attend school. Performance criteria were employed. The laterality quotient was lower among normal controls, thus indicating a preference of left-handedness among the test subjects. Greater left-handedness was more prevalent among the younger test subjects; there was no age distinction among the controls, nor between mongols and non-mongols. Males tended toward greater left-handedness than did females. (8 refs.) - *E. Kravitz.*

Bedford College
London NW1, England

2116 **MULLER, PAUL F.; CAMPBELL, H. EDWIN; GRAHAM, WILLIAM E.; BRITTAIN, HARRY; FITZGERALD, JOSEPH A.; HOGAN, MICHAEL A.; MULLER, VICTOR H.; & RITTENHOUSE, A. H.** Perinatal factors and their relationship to mental retardation and other parameters of development. *American Journal of Obstetrics and Gynecology,* 109(8):1205-1209, discussion 1209-1210, 1971.

Comparison of perinatal experience with 12 developmental parameters revealed significant relationships between the 4 perinatal factors and 6 developmental areas. This retrospective study compared data from the obstetrical charts of 1,698 children with developmental information obtained at age 9 years. Advanced maternal age was significantly related ($p<.01$) to incidence of MR and to speech defects. Breech presentation and premature rupture of the membranes were significantly related to grades repeated and lower achievement level in arithmetic, and breech presentation was significantly related to increased recommendation of remedial measures. Children who were premature or delivered with regional anesthesia had significantly lower mean scores on the Lorge-Thorndike II. Duration of anesthesia did not have significant effects. (14 refs.) - *J. K. Wyatt.*

St. Vincent's Hospital
Indianapolis, Indiana

2117 VAN UDEN, A.M.J. Congenital deafness and disturbed psychomotor development. In: Stoelinga, G.B.A.; & Van Der Werff Ten Bosch, J. J., eds. *Normal and Abnormal Development of Brain and Behaviour.* Baltimore, The Williams & Wilkins Co., 1971, p. 250-256.

Congenital deafness, defined cybernetically, is a loss of hearing in which a S cannot, without a hearing aid, hear his own voice and other sounds from his own body. Children with such deafness show retardation in eurhythmia, a condition that correlates with eupraxia, the ability to find arms, legs, and fingers. From this comes difficulty in teaching young deaf children to lipread and talk, as well as in auditory training. By means of speech and language there is, apparently, some connection between motor and cognitive development, requiring special programs of basic training for deaf children in eurhythmia, eupraxia, serial successive memory, integration of motor behavior, and movement and symbol. However, many well-educated deaf children have attained good results with respect to the purely oral way of communication and thinking. (16 refs.) - *B. Berman.*

2118 GERMAN, JAMES; EHLERS, KATHRYN H.; KOWAL, ARETA; DE GEORGE, FRANCES V.; ENGLE, MARY ALLEN; & PASSARGE, EBERHARD. Possible teratogenicity of trimethadione and paramethadione. *Lancet,* 2(7666):261-262, 1970. (Letter)

The cause of familial clustering of congenital anomalies may be seizure control drugs as well as genetic factors. Two previously described families were followed in which the mothers had petitmal seizures due to head injury and infection. During 9 pregnancies, which ended in either birth of an abnormal child or spontaneous abortion, the Ss took trimethadione or paramethadione. Withdrawal of the drugs resulted in subsequent normal pregnancies. Although developmental abnormalities did not constitute a recognizable syndrome, certain defects did recur, such as intrauterine growth retardation, cardiac malformations, clefts of the palate or lip and palate, and facial dysmorphia. Two affected surviving children are mentally retarded. Both mothers had poorer pregnancy records than normal mothers and even poorer pregnancy records than other epileptic mothers reviewed. The evidence points to the teratogenicity for man of the oxazolidine-2,4-diones, but the problem requires more data and research. (3 refs.) - *S. Glinsky.*

Cornell University Medical Center
New York, New York 10021

2119 BARMEYER, GEORGE H. Magic, science, and head circumference. *Rocky Mountain Medical Journal,* 68(5):42-44, 1971.

Adolescents with a head circumference below the third percentile have a 20:1 chance of mental deficiency. Of 1,775 Ss with a head circumference below the third percentile, only 11 (0.62%) were normal in intelligence. Among 34 adolescents with growth failure but normal intelligence, 15% were microcephalic. The use of paper tapes does not introduce significant error into head circumference measurement. Microcephaly was present in 11% of retarded outpatients. (4 refs.) - *S. Glinsky.*

501 West Broadway
Missoula, Montana 59801

DEVELOPMENTAL ASPECTS – Physical

2120 **CLAUSEN, JOHS; & KARRER, RATHE.** Autonomic activity during rest in normal and mentally deficient subjects. *American Journal of Mental Deficiency*, 75(3):361-370, 1970.

The resting levels and reactivity scores on autonomic variables in MRs tend to reflect a permanent condition rather than a temporary response to a recording situation. Analysis was made of autonomic activity in both MRs and nonretarded to test whether a possibly reduced activity in MRs might be due to an impaired arousal mechanism. The Ss were three groups of 10 each, composed of normals, nonorganically deficient MRs, and organically deficient MRs, equally divided over a CA range of 11 to 20 years. All three groups were matched for sex and the two MR groups for MA and IQ. On two occasions resting data were obtained for systolic blood pressure, blood volume changes in the head and finger, galvanic skin resistance, heart rate, heart rate variability, and respiration amplitude and period. The data indicated higher blood pressure, lower skin resistance, and a faster heart rate in the MRs. The findings contrasted with previously found decreased autonomic reactivity in MRs and indicated a more complex relationship between the arousal mechanism and autonomic activity. (17 refs.) - *S. Glinsky.*

New York State Institute for Basic
Research in Mental Retardation
Staten Island, New York 10314

2121 **LOBB, HAROLD.** Frequency vs. magnitude of GSR in comparisons of retarded and nonretarded groups. *American Journal of Mental Deficiency*, 75(3):336-340, 1970.

The retardate's rate of galvanic skin response (GSR) habituation in general is not appreciably slower than that of the nonretarded S. The GSR of 160 MR adults and 160 nonretarded adults from a prior study were reanalyzed in terms of separate frequency and amplitude measures. No evidence was found to support the hypothesis that GSR persistence is generally greater in retardate than in nonretardate groups. Actually MRs' responses were characteristically smaller and less frequent, except in extinction of conditioned responses. Although frequency data corresponded closely with earlier findings based on GSR magnitude, the magnitude analyses displayed effects of independent variables more reliably. However, these data do not necessarily refute an "inhibition deficit" concept. (10 refs.) - *S. Glinsky.*

University of Western Ontario
London 72, Ontario, Canada

2122 **RUNDLE, A. T.; & SYLVESTER, P. E.** The influence of retarded maturation on growth abnormalities in the mentally defective girl. *Journal of Mental Deficiency Research*, 14(Part 3):196-204, 1970.

Anthropometric and maturational data on 87 MR females aged 7 to 20 years were obtained to assess the effect of delay in maturation on linear growth. Measurement showed that a large proportion of the Ss were small in comparison with their normal contemporaries. When late maturation was defined as menarche after 14.4 years, results indicated that in both the early and late maturing groups there were factors predisposing the Ss towards height reduction, the additional factor of delayed puberty being significant among late maturing Ss. Delay in maturation is associated in many cases with retarded linear growth common in MRs, but some children who may be small appear to mature at the normal rate. Conversely, there are examples of normal growth patterns in females with late maturation. In any analysis of growth and maturation, the researcher must be aware of group selection errors, mechanical measurement errors, the tendency to eliminate difficult SMR Ss, and, in this case, the designation of the appropriate menarche. (23 refs.) - *B. J. Grylack.*

St. Lawrence's Hospital
Caterham, Surrey, England

2123 **FEDRICK, JEAN.** The kinetics of growth. *Lancet,* 1(7690):133-134, 1971. (Letter)

Doubt has been cast on the validity of using an infant's skull circumference as an index of the size of his brain. Analysis of 350 infant brain weight and skull circumference measurements taken during the 1958 British Perinatal Mortality Survey (excluding cases with major congenital malformations) showed a significant ($p<0.0001$) positive correlation of 0.91. It can therefore be concluded that, at birth at least, the skull circumference is closely correlated with brain weight. (1 ref.) - *J. C. Moody.*

University College
London, England

2124 **ELY, DAVID J.** A study of voluntary control of breathing in mentally retarded individuals. *California Mental Health Research Digest,* 9(1):37-38, 1971.

A definite relationship is seen to exist between voluntary breath control and speech development in MRs. Fifty-seven MR children and adolescents were preliminarily evaluated in terms of speech development and then rated as to their ability in blowing soap bubbles, whistles, and candle flames. Blowing behavior of 31 of the Ss was monitored on an oscillograph. The results showed that Ss with high and medium speech scores were able to blow with more force and duration than the low scorers. A previously unreported phenomenon consisting of a complex wave pattern in the breathing record at the onset of a blowing response was also observed. The wave pattern occurred most often in Ss who had scored better on the speech development tests. (No refs.) - *S. Glinsky.*

Porterville State Hospital
California

DEVELOPMENTAL ASPECTS – Mental

2125 **CZUDNER, GAD; & ROURKE, BYRON F.** Simple reaction time in brain-damaged and normal children under regular and irregular preparatory interval conditions. *Perceptual and Motor Skills,* 31(3):767-773, 1970.

A direct relationship between latency and length of preparatory interval was found for normal but not for brain-damaged Ss on a reaction time task. Of 15 brain-damaged Ss (CA range 8-14 yrs; IQ range 89-121), 14 had positive EEGs. Fifteen control group Ss were matched for age and IQ and had no detectable motor or sensory impairment. All Ss received training under regular and irregular preparatory interval conditions. The overall performance of brain-damaged Ss was slightly inferior ($p<.10$) than that of control group Ss and was slightly better ($p<.10$) under the regular procedures condition than under the irregular procedures condition. A separate analysis of the performance of 8 to 10-year-old Ss suggested that brain-damaged children in this age group had difficulty developing and maintaining a set. Older brain-damaged children did not demonstrate this deficit, a finding which suggests that the older group may have learned to adapt and/or recover from deficits related to inability to develop and maintain attention. (18 refs.) - *J. K. Wyatt.*

University of Windsor
Windsor, Ontario, Canada

2126 **MOSKOWITZ, HERBERT; & LOHMANN, WERNER.** Auditory threshold for evoking an orienting reflex in mongoloid patients. *Perceptual and Motor Skills,* 31(3):879-882, 1970.

Mongoloid Ss required a significantly ($p<.0005$) higher threshold (between 30 and 40 db) for a novel stimulus to evoke an orienting reflex than normal Ss. Ss were 15 institutionalized male MRs with Down's syndrome (CA range 18-35 yrs; IQ range 12-39) and 15 normal males (CA range 18-35 yrs). All Ss had normal hearing. An orienting response consisted of a galvanic skin response

(GSR) of at least 2,000 ohms or a vasoconstrictive change in a finger which was at least as great as that of the cardiac activity. A series of 1,000-cps tone bursts of increasing intensity were presented until an orienting reflex was evoked. The character of the orienting reflex differed between the groups. Two-thirds of the mongoloid Ss presented both GSR and vasoconstriction, while 8 normals presented GSR alone. These data suggest that apparent learning deficit in mongoloid MRs may be due to limitations in attention processes rather than in associative processes. (4 refs.) - *J. K. Wyatt.*

California State College at Los Angeles
Los Angeles, California

2127 **McCALL, ROBERT B.** Intelligence quotient pattern over age: Comparisons among siblings and parent-child pairs. *Science*, 170(3958):644-648, 1970.

Thirteen father-son, 11 mother-daughter, and 11 cross-sexed parent-child pairs participated in a study of the relative heritability of the general level of IQ as opposed to the pattern of IQ change over age. The parent-child comparisons were between the IQ's of the parent as a child assessed between 3 and 12 years of age and the IQ's of his or her child assessed years later when the child was 3 to 12 years of age. The results indicated a greater similarity among related siblings relative to matched controls for general level than for pattern of IQ over age. The data suggest that relatively simple trends in IQ over age occur in a large number of normal children. The genetic heritage of middle class normal children partially governs the general segment of the total IQ dimension within which an individual is likely to function, but other factors involving environmental, personality, and social interactions may determine meaningful fluctuations in mental performance over age within the general range. (14 refs.) - *B. J. Grylack.*

Fels Research Institute
Yellow Springs, Ohio 45387

2128 **SOMMERS, PAUL A.; JOINER, LEE M.; HOLT, LAURENCE E.; & GROSS, J. C.** Reaction time, agility, equilibrium, and kinesio-perceptual matching as predictors of intelligence. *Perceptual and Motor Skills*, 31(2):460-462, 1970.

Sixty-seven boys and 53 girls selected randomly from a residential population of MR children with CA of 8 to 20 years and IQ from 40 to 80 were given a kinesio-perceptual test battery in order to judge the predictive accuracy of 5 items, including dynamic equilibrium, reaction time, agility, static equilibrium, and kinesio-perceptual matching. The obtained results indicated that knowledge of the selected kinesio-perceptual test items adds 57% to the total variance accounted for by CA and sex in predicting IQ. The examined items showed low correlations with CA, indicating low developmental validity, but high correlations with IQ, emphasizing high predictive validity on an intellective criterion. Therefore, the selected kinesio-perceptual items may be used to supplement a diagnostic battery and thereby to provide a different type of measurement that might possess some direct remedial implications. (2 refs.) - *B. J. Grylack.*

Southern Illinois University
Carbondale, Illinois

2129 **SANDLER, LOUISE; VANCAMPEN, JACK; RATNER, GERALD; STAFFORD, CALVIN; & WEISMAR, RICHARD.** Responses of urban preschool children to a developmental screening test. *Journal of Pediatrics*, 77(5):775-781, 1970.

A group of 104 disadvantaged urban preschool children (ages 4 to 6) was given the Denver Developmental Screening Test in order to compare their performance with previously established norms. Both gross motor functioning and fine motor adaptive skills were comparable to Denver norms. The marked delay in acquiring language and cognitive functions reflected cultural deprivation. This lag results in a lower normative function in disadvantaged areas and points up a need for environmental enrichment rather than individual concern. Care should be used in applying middle-class norms to poverty populations. (12 refs.) - *E. L. Rowan.*

1417 Race St.
Philadelphia, Pennsylvania 19102

2130 **NEEMAN, RENATE L.; & PHILLIPS, HERBERT E.** Perceptual-motor survey of young adult retardates. *Perceptual and Motor Skills*, 31(3):1000, 1970.

Significant test-retest correlations on the Purdue Perceptual-motor Survey may reflect the high frequency of perceptual-motor dysfunction among MRs and may imply dysfunction syndromes. The survey was administered, an average of 59 days apart, to 36 MRs (mean age 23.3 yrs; mean IQ 52.9; 52.8% males; 22.2% mongoloid). Significant test-retest correlations ($p<.01$) were found for all items except the Chalkboard: Double Circle test. Items with the highest correlations included Ocular pursuits of the right eye, the left eye, and both eyes; Rhythmic Writing; and Walking Board. Walking Board and Jumping were correlated. Imitation of Movement was correlated with Walking Board:Backward and Jumping. IQ was correlated with items 5 and 6; 23 and 24; 25 and 26; and 37 and 38 at a significant level ($p<.01$). The relationship between sex and items 9 and 10 was significant ($p<.01$). (2 refs.) - *J. K. Wyatt.*

State University of New York
Buffalo, New York

2131 **PRENTICE, NORMAN M.; & BIERI, JAMES.** *Intellectual development of culturally deprived children in a day care program: A follow-up study.* Presented at 47th Annual Meeting, American Orthopsychiatric Association, San Francisco, March 25, 1970. (13 pages, mimeo)

In an effort to determine the effects of day care on intellectual development, retesting with Stanford-Binet and Goodenough-Harris, after 9 months, of 2 groups of culturally deprived day care children revealed that those who had been in the day care program an average of 23 months ("old" children) showed no significant difference on the G-H test, but exhibited a slight drop on the Binet; those in the program an average of 12 months ("new" children) showed statistically significant gains on both tests, especially the G-H. For the "new" children, the 9 additional months of day care reflected improvement in the child's competence in tasks comparable to those performed in everyday school activities. Failure of the "old" children to show further development suggests that culturally deprived children in a traditional preschool program often reach a plateau in their second year; only radical innovations, it is suggested, will stimulate further intellectual growth. Unanswered questions include: What factors in day care experiences contribute to cognitive development? Are some curricula more effective than others? What are the long-range effects? (12 refs.) - *B. Berman.*

University of Texas
Austin, Texas

2132 **LUCAS, MARILYN S.** Assessment of coding behavior of trainable retardates. *American Journal of Mental Deficiency*, 75(3):309-315, 1970.

Visual presentations are more effective in facilitating recognition in MR than either cross-modal or auditory presentations. The differential effects of cross-modal and unimodal coding, and preferred-sense modality upon ability to recognize previously presented stimuli, were studied. The Ss were 96 institutionalized MRs with CAs between 10 and 20 years and IQs between 35 and 59. They were given 6 subtests of the Illinois Test of Psycholinguistic Abilities (ITPA) and divided into High Auditory and High Visual modality groups. Each group performed a recognition task under unimodal (visual-visual or auditory-auditory) or cross-modal (visual-auditory or auditory-visual) conditions of presentation and recognition. The results indicated no relationship between assessed modality strength on the ITPA and performance on the recognition task. The visual presentation-visual recognition task condition exhibited significantly higher scores than the cross-modality or auditory-auditory task conditions. The evidence indicates that visual presentation is clearly superior to other modality presentation. (16 refs.) - *S. Glinsky.*

University of California, Riverside
Riverside, California 92502

2133 **WRIGHT, LOGAN.** Prerest performance levels of nonretardates and retardates, and reminiscence on a manual crank test. *American Journal of Mental Deficiency*, 75(3):304-308, 1970.

Significant differences favoring nonretardates on reminiscence scores existed where prerest performance was not controlled, while no reminiscence

differences were found when prerest performance was controlled. The discrepant findings regarding the relationship between reminiscence and reactive inhibition (IR) on one hand, and MR on the other hand, were investigated. The Ss were 36 male MRs with IQs between 61 and 78, and 36 nonretarded Ss with IQs between 90 and 110, all of whom performed 20 10-second manual crank trials interrupted with 10-second inter-trial intervals. After the 20th trial the Ss were given a 3 minute rest, followed by 10 postrest trials. The rate of prerest performance for subgroups of 16 MRs and 16 nonretarded was controlled. Two other subgroups had no control over their cranking rate during the prerest period. In the second group over which no control was exerted nonretardates performed at a higher prerest level and showed more reminiscence. No differences were found between the reminiscence scores of MRs and nonretardates where the level of prerest performance was controlled. These results suggest that motivational factors cause nonretardates to perform at a higher level of prerest performance and thus build up greater amounts of reactive inhibition. (12 refs.) - *S. Glinsky.*

University of Oklahoma Medical Center
Oklahoma City, Oklahoma 73104

2134 **DAS, J. P.** Vigilance and verbal conditioning in the mildly and severely retarded. *American Journal of Mental Deficiency*, 75(3):253-259, 1970.

Vigilance and verbal conditioning tasks given to MRs examined the orienting response as the probable link between learning and attention. The institutionalized Ss consisted of one group of EMRs with a mean MA of 113.21 months and mean IQ of 66.10, and a second group of SMRs with a mean MA of 84.96 months and mean IQ of 47.85. Both groups had mean CAs of 169 and 177 months, respectively. The Ss performed a verbal conditioning task consisting of acquisition and reversal phases, and a simple and a complex vigilance task. The results showed that the SMRs were slower in response speed than the EMRs in conditioning, which was attributed to the formers' relative immobility of nervous processes. In simple vigilance both groups exhibited similar trends in decrement as a function of time at task. In complex vigilance the SMRs were inferior to the EMRs. A direct variation between proficiency in vigilance and conditioning performed was noted upon comparing good and poor performers in the one vigilance task on performance and in the other vigilance task and in conditioning. The results suggest that the orienting response is the basic connection between vigilance and conditioning. (22 refs.) - *S. Glinsky.*

University of Alberta,
Alberta, Edmonton 7, Canada

2135 **DREW, CLIFFORD J.; & PREHM, HERBERT J.** Retention in retarded and nonretarded children as a function of direction of recall and material associative strength. *American Journal of Mental Deficiency*, 75(3):349-353, 1970.

Material associative strength influences recall performances of MRs and nonretardates to the same relative degree, while direction of recall more greatly affects MR performance. The recall performance of MRs and nonretardates was compared as a function of direction of recall task and material associative strength. The Ss were 48 MRs with IQs between 55 and 72 and 48 Ss with IQs between 100 and 115, all sampled randomly from a public school pool with CAs between 10 years and 12 years, 4 months. The Ss learned two lists of 14 word-pairs selected from Palermo and Jenkins and gauged according to immediate, 24-hour, and 15-day recall performances. The results indicated significantly fewer correct responses by MRs after 24 hours and 15 days, but not on immediate recall. Direction of recall task influenced MR performance to a greater degree than nonretardate performance. S performance on high associative strength material was significantly better than on low associative strength material at 15-day testing only. The findings indicated that review is important for better recall and that level of training might influence response availability. (22 refs.) - *S. Glinsky.*

University of Texas
Austin, Texas 78712

2136 **GALBRAITH, GARY C.; GLIDDON, JACK B.; & BUSK, JYTTE.** Visual evoked responses in mentally retarded and nonretarded subjects. *American Journal of Mental Deficiency*, 75(3):341-348, 1970.

Reliable visual evoked response (VER) differences exist between nonretarded and MRs. The Ss were 24 institutionalized MRs with a mean CA of 16.3 years and mean IQ of 46.7, and 16 nonretarded with a mean CA of 20.0 years and undetermined IQs. The VER was recorded from scalp electrodes placed over primary visual cortex and associative cortical fields. In general the MRs showed significantly longer VER latencies, failure to develop certain VER components, and reduced amplitudes in the topographic distribution of VERs recorded over associative cortical fields. These findings give general support to several postulates of retardate neural functioning derived from Gestalt field theory. (16 refs.) - *S. Glinsky.*

Pacific State Hospital
Pomona, California 91766

2137 WYNE, MARVIN D.; COOP, RICHARD H.; & BROOKHOUSE, DIANE B. Information processing in young mildly retarded children. *American Journal of Mental Deficiency*, 75(3):371-375, 1970.

Cognitive style and conceptual tempo are viable determinants of the idiosyncratic modes which MR children utilize in processing information. The Ss were 40 EMRs (20 male, 20 female) with CAs of 8.7 to 13.11 years and MAs from 4.5 to 10.5 years. All Ss were individually administered the Sigel Cognitive Test (SCST), Matching Familiar Figures Test (MFF), and four performance subtests of the Wechsler Intelligence Scale for Children. The findings did not uphold several performance predictions, showed that conceptual tempo seemed to be a more sensitive predictor of performance than cognitive style, and implied that EMRs display a similar range in cognitive style and conceptual tempo to that of nonretardates. Also, significant sex differences in cognitive functioning were found. The data indicate the need for development of more sensitive determinants beyond IQ and achievement test responses for educational placement and programming. (12 refs.) - *S. Glinsky.*

University of North Carolina
Chapel Hill, North Carolina 27514

2138 CARR, JANET. Mental and motor development in young mongol children. *Journal of Mental Deficiency Research*, 14(Part 3):205-220, 1970.

Fifty-four mongol infants and an equal number of controls were included in a 2-year longitudinal study of mental and motor development. The Bayley Infant Scales of Mental and Motor Development were used at 5 different times during the 2 years to assess this development. In both areas of assessment, the test results of the mongol Ss fell off precipitously to 10 months of age with a more gradual drop to 2 years. The mean scores of the 45 Ss who were brought up at home were higher than those of Ss raised in foster homes and institutions. Female Ss scored better than male Ss. Mean scores on the Motor Scale were below those on the Mental Scale after 6 months of age, a trend which may not continue past 2 years of age because of the timing of the ability to walk in these children. Although the sample was too small to allow a comparison of the relative merits of institutions and foster homes for rejected mongols, it is evident that the mongol child has the best chance of developing his full potential if he is raised by his family in his own home. (6 refs.) - *B. J. Grylack.*

University of London Institute of Education
London, W.C. 1, England

2139 WILLERMAN, LEE; NAYLOR, ALFRED F.; & MYRIANTHOPOULOS, NTINOS C. Intellectual development of children from interracial matings. *Science*, 170(3964):1329-1331, 1970.

Eighty-eight children of interracial matings who were at least 4 years of age were tested with the Stanford-Binet IQ format. In assessing the postnatal effect of the race of the mother on the IQ of the 4-year-old child, the marital status of the mother and the sex of the baby were taken into account, since these factors have been found significant. The results indicate that if sex is ignored, maternal race is significant only among the children of unmarried mothers ($P<.05$), and that among Negro mothers it is the children of the unmarried who have the low IQs ($P<.05$). Alternatively, if marital status is ignored, maternal race is significant only among males ($P<.05$), and among Negro mothers it is the male children who have the low IQs ($P<.05$). Therefore, the male children of unmarried Negro mothers have the lowest IQs. The evidence suggests that

environmental factors may play an important role in the lower intellectual performance of Negro preschool children. Within the same framework, larger numbers of children will be followed. (14 refs.) - *B. J. Grylack.*

University of Michigan
Ann Arbor, Michigan 48104

2140 **WEAVER, L. A.; & RAVARIS, C.** The distribution of reaction times in mental retardates. *Journal of Mental Deficiency Research*, 14(4):295-304, 1970.

An examination of the distribution of reaction time (RT) in institutionalized MRs disclosed that a liberal upper limit for RT should not be used to make decisions regarding capabilities. A conventional RT test was administered as part of a test battery to 270 institutionalized MRs, 78 of whom were mildly retarded, 178 of whom were moderately retarded, and 14 of whom were SMR. Five usable trials were taken. When the RT was < 0.100 second, it was considered an anticipatory response, and when it exceeded 1.99 seconds, it was defined as a failure of attention. In these cases, the trial was repeated. Mean reaction times were 0.530 second for the total group, 0.376 for mildly MR Ss, 0.579 for moderately MR Ss, and 0.762 for SMR Ss. Discrepancies between the group mean and median were significantly skewed ($p<.05$) toward the higher range values. Distributions of individual means, classified in 0.1 second intervals, were significantly skewed ($p<.05$), and 85% lay below the midpoint of the score range. Distribution of standard deviations associated with means was normal. Most of the groups did not have any excessively long RT values, and the majority of responses were less than 1.0 second. A 2-second RT limit would have included virtually all trials. Very long reaction times should not be accepted as a genuine response. (7 refs.) - *J. K. Wyatt.*

University of Vermont Medical
Alumni Building
Burlington, Vermont 05401

2141 **WILLERMAN, LEE.** Biosocial influences on human development. *American Journal of Orthopsychiatry*, 42(3):452-462, 1972.

Racial and socioeconomic differences in the incidence of maternal complications during pregnancy parallel findings on mean IQ differences between races and socioeconomic groups. Data for a collaborative study in 12 hospitals were compiled for pregnant women who received prenatal care and delivered 56,000 infants who are being followed until 8 years of age. The sex ratio was significantly depressed ($p<.01$) for the lowest socioeconomic groups. The sex ratio for whites consistently exceeded that for Negroes within the same socioeconomic category. There was a positive correlation between socioeconomic index and 4-year Stanford-Binet IQ within white and Negro populations; Negro Ss averaged 8 to 13 points lower than white Ss in the same socioeconomic group. In general, high IQ and high birthweight were related. The degree of advantage associated with higher parental socioeconomic status was not as high for Negro children as for white children. Mean gestational age for Negro children was somewhat shorter than that for white children. Among the diseases identified as related to high IQ were retinoblastoma and asthma. These findings suggest the influence of a biochemical product on brain development. The source of differences between Negroes and whites may lie in the inadequate assessment of social class. Early diagnosis of defects of the nervous system may provide opportunity for environmental intervention. (32 refs.) - *J. K. Wyatt.*

University of Texas
Austin, Texas

2142 **SERAFICA, FELICISIMA C.** Object concept in deviant children. *American Journal of Orthopsychiatry*, 41(3):473-482, 1971.

In an assessment of object concept in children with a history of deviant development, the performance of 8 Ss, ranging in age from 4 to 8 years, was tested on a scale for visual pursuit and permanence of objects. The Ss had various diagnoses but manifested such common characteristics as impaired interpersonal relationships, communication difficulties, and lag in intellectual growth. Greater variability of performance was observed in this small but deviant population than among a larger group of normal infants in another study. The differential reactions to the preferred and neutral objects in the study

denoted a horizontal decalage favoring the preferred object in which the child attains a more advanced stage in the search for the preferred object as compared with the neutral one. It appears that behavior indicative of object concept attainment will be displayed generally where there is a cathectic bond or relationship as expressed in preference for an object. (28 refs.) - *B. J. Grylack.*

Clark University
Worcester, Massachusetts 01610

2143 **HERTZIG, MARGARET E.; & BIRCH, HERBERT G.** Longitudinal course of measured intelligence in preschool children of different social and ethnic backgrounds. *American Journal of Orthopsychiatry*, 41(3):416-426, 1971.

Sixty Puerto Rican children and 116 white middle-class children were examined at 3 years of age, and 57 and 110 children, respectively, of these groups were reexamined at age 6 with the Stanford-Binet, Form L, in order to determine the stability and change in IQ of these children from preschool to school age. Mean intelligence test scores at both ages provide no indication of a decline in measured intelligence in the Puerto Rican children. Both the middle-class and Puerto Rican groups show a trend toward higher IQ at the later age, with the group increment being significant in the middle-class group and approaching significance in the Puerto Rican group. These findings are at variance with a frequently expressed opinion that children at a social disadvantage show declines in measured intelligence with age during the preschool years. Although the degree of disadvantage may have been insufficient in this sample, such an interpretation is unlikely in view of the agreement of the results with other studies. (17 refs.) - *B. J. Grylack.*

New York University School of Medicine
New York, New York 10016

2144 **O'GRADY, DONALD J.; BERRY, HELEN K.; & SUTHERLAND, BETTY S.** Cognitive development in early treated phenylketonuria. *American Journal of Diseases of Children*, 121(1):20-23, 1971.

Phenylketonuric children treated early in the course of the disease did not exhibit any deficit in total psycholinguistic abilities, with the exception of one significant deficit in specific functioning. Eight phenylketonuric children receiving early treatment, all over 3 years old, and their 6 siblings were evaluated with the Stanford-Binet Intelligence Scale and the Illinois Test of Psycholinguistic Abilities (ITPA). IQ scores and ITPA total and subtest scores were analyzed with the Mann-Whitney U test. In contrast to some reports, those children treated early did not differ significantly from their siblings on either the Binet IQ or the ITPA total score. A significant difference was found on the ITPA subtest of vocal encoding, which may be due to small sample size. The ITPA profiles from repeated testing in 4 cases were not stable, and observed fluctuations did not appear related to any observable changes in intellectual or academic functioning. Since the IQ measure as a broad measure can obscure specific behavioral deficits, the search for more sensitive measures is warranted. (24 refs.) - *S. Glinsky.*

Children's Hospital Research Foundation
Cincinnati, Ohio 45229

2145 **POLLITT, ERNESTO.** Poverty and malnutrition: Cumulative effect on intellectual development. *Les Carnets de l'Enfance/Assignment Children*, 14:40-52, 1971.

The general relationship between low intellectual performance and low social class has been well established. Within the limits set by genotype, the rate of intellectual development of a child during the formative years will partly depend on the nature of the social and biological environment within which the child develops. Factors from both sets of environments can hinder cognitive growth by either restricting or not making available information required for the development of mental abilities or by directly affecting the organism so that it cannot utilize or absorb the available information. Aside from adverse social and biological conditions prevalent among low income groups, such as insufficient learning opportunities, the malnourished child could suffer the effects of 2 other sets of conditions, specifically those intimately related to the nutritional deficiency, such as non-responsiveness to environmental stimuli because of malnutrition-induced apathy, and the nutritional deficiencies

per se. While it will not be possible in the foreseeable future to assess precisely the contribution of each of the factors involved in significantly limiting the cognitive growth of the child, any program aimed at the rehabilitation or prevention of intellectual deficits must consider the overall medical and social conditions. (55 refs.) - *P. Custers.*

United Nations Research Institute
for Social Development
Geneva, Switzerland

2146 GOLDHAMER, HERBERT. Children of interracial matings. *Science*, 172(3978):10, 1971. (Letter)

Differences in IQs between children of Negro father-white mother couples and white father-Negro mother couples suggest that the mean IQ of the first group of couples is higher than that of the second. A study of Boston intermarriages has shown that Negro husbands were well above Negro males in occupational attainment, whereas white husbands were far below the white male. This could indicate an absolute superiority in intelligence of the Negro husbands over the white husbands. Since there is no reason to suppose that Willerman's couples are different from the Boston couples, and since the educational attainments of the Negro fathers are superior to those of the white, it is reasonable to assume different mean IQs in the two types of couples studied. (1 ref.) - *N. Jarvis.*

Rand Corporation
Santa Monica, California 90406

2147 WALBERG, HERBERT J. Children of interracial matings. *Science*, 172(3978):8, 1971. (Letter)

The observation by Willerman et al. that interracial children with white mothers scored higher on the Stanford-Binet IQ test than did interracial children with Negro mothers led the observers to infer that environmental factors depress the intellectual performance of Negro children. This inference cannot be allowed, because the observers did not test the effect of maternal race independently, but combined it with the effects of the sex of the child and the mother's marital status. The question whether the mother's race has a significant independent effect cannot be answered with this kind of analysis. Moreover, the simple regression slope cannot be interpreted for each of the 3 effects, since each effect is confounded with the other 2. Two-way interactions involving maternal race, in 2 analyses leaving out sex and marital race, respectively, may be accounted for by a confounding of effects of the excluded factor and maternal race. (3 refs.) - *N. Jarvis.*

University of Illinois
Chicago, Illinois 60680

2148 HENDERSON, NORMAN D. Children of interracial matings. *Science*, 172(3978):8-9, 1971. (Letter)

A 7-point difference in IQ scores of offspring of interracial marriages, reported by Willerman et al., may be explained by paternal genetic differences rather than maternal environmental effect. Among their 4 types of parents, the Negro fathers had the highest educational level, but ordinarily this group is characterized by the lowest level of educational attainment. This suggests that Willerman's group of Negro parents may have been more intellectually gifted than the other parents. Data on the intelligence levels of the fathers in this study might suggest a different interpretation of influences on the children's IQs. (1 ref.) - *N. Jarvis.*

Oberlin College
Oberlin, Ohio 44047

2149 VAN DEN DAELE, LELAND. Children of interracial matings. *Science*, 172(3978):10, 1971. (Letter)

In a study of interracial matings, self-selection factors must be taken into consideration in inferences from data. Negro males who mate with white women may be characterized by intellectual and motivational factors which are reproduced in the children of such unions. But the fact that the male children of Negro mothers had the low IQs in Willerman's study suggests that a race-related, sex-linked control of developmental processes related to intelligence may be involved. The observers have assumed that additive genetic

factors are not sex-linked, but they do not hesitate to invoke sex linkage when environment fails as a sufficient cause for sex difference. The possibility of alternate explanations of Willerman's findings indicates that environmental interpretations of race effects and interactions should not be made prematurely. (7 refs.) - *N. Jarvis.*

University of Illinois
Urbana, Illinois 61801

2150 WILLERMAN, LEE; NAYLOR, ALFRED F.; & MYRIANTHOPOULOS, NTINOS C. Children of interracial matings. *Science*, 172(3978):12, 1971. (Letter)

In reply to objections: 1) Our table 2 allows maternal race effect to be tested controlling for sex of child and marital status. Race-of-mother effect is absent only for female children of married mothers. 2) Henderson pools educational attainment over all age groups, concealing the fact that education is confounded with age. Data from our Table 1 are contrary to Henderson's assertions, nor is his point about the ceiling effect substantiated. 3) Goldhamer's data of 30 years ago are not comparable; moreover, his data do not support his claim. 4) Genetic explanations of our data would require that loci affecting a quantitative character all be on one of the 23 haploid chromosomes, which seems unreasonable. Moreover, we did not invoke sex-linkage to explain the observed sex difference. X chromosome loss is not to be identified with sex linkage. (2 refs.) - *N. Jarvis.*

University of Michigan
Ann Arbor, Michigan 48104

2151 STOELINGA, G.B.A.; & VAN DER WERFF TEN BOSCH, J. J., eds. *Normal and Abnormal Development of Brain and Behaviour.* Baltimore, The Williams and Wilkins Co., 1971, 348p.

Presenting the contents of a postgraduate course for medical practitioners of various specialties, this volume includes concrete information on the development of brain and behavior, and on the influence of various internal and environmental factors on neural development. Providing insight into methodological parameters pertaining to the study of behavioral growth, the volume also furnishes clues to defining and scoring behavioral items and identifying separate elements in a particular behavior. Held in Leiden, Holland, in November 1971, the course was part of a series initiated by the Dutch Growth Foundation. (550 refs.) - *B. Berman.*

CONTENTS: Somatic Development and the Study of the Central Nervous System (Marshall); Undernutrition and the Developing Brain (Dobbing); Effects of Early Malnutrition on General Development in Animals (Widdowson); Postnatal Growth and Development in Small-for-Date Babies (van Gils); On the Structure, Development, and Connections of the Limbic System (Kappers); Behavioral Assessment in Infancy (Freedman); Manual to Accompany Newborn Behavioral and Neurological Scales (Brazelton and Freedman); Motor Behaviour in Relation to Brain Structure (Prechtl); Neurological Criteria for the Estimation of the Post-Menstrual Age of Newborn Infants (Casaer and Akiyama); Neurological Follow-up of Infants Born After Obstetrical Complications (Touwen); Observations of Free-Field Behavior in Preschool Boys and Girls in Relation to Neurological Findings (Kalverboer); Genetic Influences on Development of Behavior (Freedman); Non-Genetic Prenatal Influences on Psychological Development (Jans); Congenital Deafness and Disturbed Psychomotor Development (van Uden); Effects of Congenital Blindness on Development of Behaviour (van Hof); Influence of Emotional Deprivation on Growth and Behaviour (Croughs); Neuroendocrine Factors and the Ontogeny of Behavior (Levine); The Effects of Early Hypothyroidism on IQ, School Performance, and Electroencephalogram Pattern in Children (van Gemund and de Angulo); Pituitary-Adrenal Hormones and Behaviour (Wied); Hormonal and Social Determinants of Sexual Behaviour in the Pigtail Monkey (Goldfoot).

2152 LEICHT, KENNETH L. *Implicit Associations in Normals and Retardates. Project Summary.* Normal, Illinois, Illinois State University, 1971, 16p.

The combined effects of high or low meaningfulness and high or low orthographic similarity with paired associate stimuli on normal and MR Ss were compared to investigate the fruitfulness of defining encoding differences in terms of differences in ability to produce implicit verbal associations. Encoding processes were viewed as implicit verbal associations. It was presumed that

normal Ss would be more likely to encode verbal units spontaneously as implicit associative responses. If this were correct, orthographic similarity would have a detrimental effect for low meaningful stimuli in normal Ss and for low and high meaningful stimuli in MR Ss. (No refs.) - *J. K. Wyatt.*

Illinois State University
Normal, Illinois 61761

2153 **ROSS, ROBERT T.** IQ changes in hospitalized retardates. *California Mental Health Research Digest*, 9(1):30-31, 1971.

Decreases in IQ in hospitalized MRs are fictitious and derive mainly from increases in the patients' chronological ages. The Ss were 324 MR patients in Fairview State Hospital in California. Their IQs were determined on admission and again at a later date and then used in an analysis of hospital age (HA), change in IQ, and change in MA. The results showed a decrease in IQ as HA increases, larger IQ decreases in those with higher IQs and lower CAs at admission, and no relationship of MA changes to HA. IQ is thus inversely related to CA when the first IQ measurement is made in childhood. MA taken alone seems to be a more meaningful measure. (No refs.) - *S. Glinsky.*

Fairview State Hospital
California

2154 **TOUCHETTE, PAUL E.** Transfer of stimulus control: Measuring the moment of transfer. *Journal of the Experimental Analysis of Behavior*, 15(3):347-354, 1971.

SMRs were able to acquire simple form discriminations without error. The Ss were 3 SMRs in whom the moment of stimulus control transfer was investigated. Each was first taught to press a red key versus a white key simultaneously present. After this discrimination had been established black figures were superimposed on the red and white keys. Each correct response affected the next trial by delaying the onset of the red stimulus an additional 0.5 second. Transfer of stimulus control to the figures was indicated when the Ss responded correctly before the onset of the red stimulus. A series of errorless discrimination reversals was accomplished with this technique, during which the number of trials to transfer systematically decreased with successive reversals. (19 refs.) - *S. Glinsky.*

E. K. Shriver Center
Waltham, Massachusetts 02154

2155 **MacKAY, C. K.; & BROWN, JEAN.** A learning strategy in normal children and severely subnormal adults. *Psychonomic Science*, 23(6):391-392, 1971.

Normals and SMRs do not appear to use different learning mechanisms in simple color sorting discrimination. Bryant (1967) suggested that normals employ an "A/not A" strategy, while SMRs learn the 2 responses relatively independently. In a modification of Bryant's experiment, an initial training task, in which groups of normal children and SMR adults, matched on MA, learn to sort black and white cards, is followed by 1 or 2 transfer tasks. One consists in sorting black and red cards and the other in sorting white and red cards. No support is found for Bryant's view. (3 refs.) - *S. Glinsky.*

University of Aberdeen
Aberdeen AB9 2UB, Scotland

2156 **WARD, L. CHARLES; & SIEGEL, PAUL S.** Effects of delay of reinforcement and stimulus meaningfulness on the discrimination learning of retardates at two intelligence levels. *Psychonomic Science*, 23(6):395-396, 1971.

The effects of delay of reinforcement and stimulus meaningfulness upon the discrimination learning of MRs at 2 intelligence levels were reliable but not significant. Ss from a borderline and mild MR group learned a left or right motor response to each of 6 pictures, 3 meaningful and 3 nonmeaningful. For a given S, each stimulus from both meaningfulness categories was paired with a different delay interval of 0, 3, or 9 seconds. The results indicated reliability of the effects, but none of the interactions reached significance. (12 refs.) - *S. Glinsky.*

Murray State University
Murray, Kentucky 42071

DEVELOPMENTAL ASPECTS – Social and Emotional

2157 **FOREHAND, REX; & BAUMEISTER, ALFRED A.** Effect of frustration on stereotyped body rocking: Follow-up. *Perceptual and Motor Skills,* 31(3):894, 1970.

Frustration significantly ($p<.05$) increased the stereotypy rate of body-rocking in 8 institutionalized male SMRs. All Ss engaged in frequent body-rocking. Frustration was induced by abruptly withdrawing an edible substance before consumption was completed. The effects of frustration were observed for 2 days in one experiment, but were not apparent after the first day in a second experiment. (No refs.) - *J. K. Wyatt.*

University of Oregon
Portland, Oregon

2158 **STERNLICHT, MANNY; PUSTEL, GABRIEL; & DEUTSCH, MARTIN R.** Suicidal tendencies among institutionalized retardates. *Journal of Mental Subnormality,* 16(31):93-102, 1970.

Identification of 45 male and female suicidal MRs in an institutionalized population indicated an incidence of 1.0% for males, 0.5% for females, 0.4% for adolescents, 2.0% for adults, and 0.9% for the total suicidal population, as compared with the total institutional population. Mean IQ for suicidal MRs was within the mildly retarded range. Suicidal females had slightly higher IQs than suicidal males, and adolescents tended to have higher IQs than adults. Average duration of institutionalization was 18 years, with a range of from 5 to 52 years. The primary medical diagnostic category for the group was familial MR. The group had a history of frequent visits by relatives and friends. Suicidal residents tended to come from broken homes and from families where they had encountered rejection or neglect. Among the prominent behavioral characteristics displayed by the group were depression, aggression, moodiness, destructiveness, outbursts of temper, psychotic and schizophrenic symptoms, emotional hypersensitivity, feelings of being rejected, and homosexual inclinations. A theory of suicidal behavior in the MR drawn from these data states that suicide is seen by the MR as the one possible remaining act in a life which has been filled with psychological misfortunes. Suicide is used both as an escape from an intolerable and hated reality and as means of gaining revenge. If the lives of the MR were less protected and less structured, the incidence of attempted suicide would probably increase. (25 refs.) - *J. K. Wyatt.*

Willowbrook State School
Staten Island, New York

2159 **WOLLEN, W.** Psychiatric illness among the subnormal. *Nursing Mirror,* 131(10):28-31, 1970.

Efforts to identify certain cases of mental subnormality with childhood neurosis or psychosis tend to blur distinctions between various forms of developmental behavioral disorder and may lead to some confusion. Before the eighteenth century, there was an amazing lack of reference to emotional disturbances in children, and until 1940 psychoses were diagnosed only rarely in childhood. Subsequent research on psychiatric disorders in the mentally subnormal, including schizophrenia, hypomania, psychoneuroses, and dissociative disorders, has shown that many questions remain in regard to the association between psychoses, psychoneuroses, and mental subnormality. The main difficulty is that most definitions of mental disorder carry the implication of homogeneity and have involved researchers in numerous assumptions, many of which are invalid. Despite endless uncertainties, many practical and scientific findings point to significant developments in the area of psychiatric disorders among the subnormal in the future. (6-item bibliog.) - *B. J. Grylack.*

Darenth Park Hospital
Dartford, Kent, England

2160 **CALDWELL, BETTYE M.** The effects of psychosocial deprivation on human development in infancy. *Mental Health Digest,* 2(12):23-28, 1970.

Studies of the effects of psychosocial deprivation on the human infant have resulted in more speculation than investigation. Available studies are consistent in implying that infants from deprived and non-deprived homes appear to develop at approximately the same rate during the first year of life, but the period from age 1 to 3 has been neglected. By age 3, the deficit of the deprived child is striking and resistant to sustained change. The findings suggest that the optimal time for an attempt at reversing deprivation is during the 2-year hiatus when Ss are seldom visible to research scrutiny. Areas requiring more attention in research of this type include improved techniques for assessing the psychosocial environment, change-sensitive measures of early child development, exploration of the relationship between constitutional factors and susceptibility to the influences of deprivation, and research utilization of the clinical single-case model. (No refs.) - *B. J. Grylack.*

No address

2161 **FRASER, WILLIAM I.** A retrospective and cross-sectional investigation of a deviant subcultural group. *American Journal of Mental Deficiency,* 75(3):298-303, 1970.

Patterns of crime among retardates who were studied years after dissolution of a therapeutic community did not differ from either prior offenses or conduct in the hospital. The relationship of criminal antecedents to prognosis of subsequent criminal behavior was investigated. The Ss were 513 behaviorally disturbed, retarded, and borderline retarded males from a Scottish therapeutic hospital who were studied retrospectively and by cross-sectional analysis 10 years after termination of the community. The CA range of the Ss was 7 to 16 years and the IQ range 45 to 85, with 249 of the Ss possessing criminal records prior to hospital admission. Records of 85% of the original group were found and analyzed, and 93 Ss were interviewed and given psychometric tests. The results indicated that for a proportion of the whole the behavior pattern which determined admission to the original hospital was unaltered by psychiatric remand or environmental therapy. There was a spectrum of behavioral symptomatology between the recidivist and nonrecidivist. This type of "follow up" approach continues to remain necessary and valid. (26 refs.) - *S. Glinsky.*

Lynebank Hospital
Dunfermline, Fife, Scotland

2162 **COLLINS, HARDIN A.; BURGER, GARY K.; & DOHERTY, DANIEL.** Self-concept of EMR and nonretarded adolescents. *American Journal of Mental Deficiency,* 75(3):285-289, 1970.

EMR adolescents tend to have more negative self-concepts than nonretarded adolescents. The self-concepts of EMR young people in a special education school were compared with those of intellectually normal youths attending a public high school. The Ss were 42 EMRs and 49 nonretarded normal youths. The measure used was the Tennessee Self-Concept Scale, composed of 100 self-report items yielding scores on 14 different scales. For the EMRs 4 scales showed no significant differences, while the remaining scales were all negative, 4 of them significantly. In addition there was a general negative perception and low esteem in both groups. Future research in this area should investigate specific dimensions of the self-concept rather than employing a single global measure. (20 refs.) - *S. Glinsky.*

Saint Louis University
St. Louis, Missouri 63103

2163 **VALLETUTTI, PETER.** Language of the mildly mentally retarded: Cognitive deficit or cultural difference? *Exceptional Children,* 37(6):455-459, 1971.

Mild MR, among individuals from lower socioeconomic backgrounds, may be partly due to cognitive confusion resulting from illogical deviant language patterns. Liberal approaches to language evaluation emphasize the relationship aspect rather than the cognitive aspect of language. The conservative approach views the linguistic code of lower class adults as ideally suited to the maintenance of social relationships but

poorly suited to the development of careful reasoning, hypothetical ideas, or complex thinking. Language evaluation should alternate between both perspectives. Educational priority should be given to the cognitive function of language. The social acceptability of language should receive only minor consideration. Those aspects of language which are critically related to the development of intelligence should be identified and distinguished from benign dialectical aspects. Teaching should emphasize those aspects of speech which are vital to thought processes; errors of language which interfere with cognition should be identified and remedied. Education should not interfere with aspects of language which do not damage cognitive development and which may facilitate intragroup communication. (35 refs.) - *J. K. Wyatt.*

Coppin State College
Baltimore, Maryland

2164 **CHATAGNON, P.-A.** Le retard de l'organisation et du developpement des fonctions mentales chez l'etre humain. I.—Retard mental et capacite sociale (Retardation of the organization and development of mental functions in the human being. I.—Mental retardation and social capacity). *Annales Medico-Psychologiques*, 2(4):497-544, 1970.

The worldwide problem of mental defects is closely connected with the problem of acquisition by MRs of a valid social capacity, which can be achieved through early recognition of the affliction, education, and participation in professional life in a form adapted to their individual qualifications. Comparison of 20 MR individuals ranging in age from 10 to 18 years showed that there are as many cases of MR alone as MR presenting in association with other concomitant pathological processes. The fact that 50% demonstrated neurological, neurotic, psychotic, and endocrine disorders related to the evolution of the mental defect is significant, for it proves that adult mental diseases are manifested frequently at a very young age. In addition to prenatal traumas, infections, and intoxications, the postnatal environment of the child has great importance. Of the 20 Ss studied, only 4 came from families in which the mother or father was normal. Any investigation of the evolution of the MR child cannot be separated from a study of his family and his milieu. (No refs.) - *B. J. Grylack.*

No address

2165 **SKABERNE, BRONISLAV.** Untersuchungen en Delinquenten Jugendlichen in Slovenien (Investigations of young delinquents in Slovenia). *Heilpadagogischen Forschung*, II(2):180-189, 1970.

Three recent research projects related to problems of youthful delinquents in Slovenia are described. The first concerns an investigation into the personality structures of 97 young male delinquents who were retarded both mentally and physically. The objective was the determination of those essential biological, psychological, and sociological factors common to most young delinquents in Slovenia. Comparison with nondelinquent students from the area revealed significant differences in level of intelligence, sexual behavior, and other factors. The second research project involved an inquiry into the social background of young Slovenian delinquents through a study of 58 families of non-delinquent young persons and an equal number of families of delinquents. Differences in housing conditions, school attendance, and parental attitudes toward education were considered, among others. The third research project investigated young persons who, because of antisocial behavior, had been placed in new families. Eighteen persons were compared with young persons who had, for the same reasons, been placed in educational institutions. The environment of the educational institutions was found to be less favorable for the child than the family situation, but the individual's specific personality structure should be considered in deciding which environment is best. (No refs.) - *P. Custers.*

Medvedova 10/11
Ljubljana, Yugoslavia

2166 **KALVERBOER, A. F.** Observations of free-field behaviour in preschool boys and girls in relation to neurological findings. In: Stoelinga, G.B.A.; & Van Der Werff Ten Bosch, J. J., eds. *Normal and Abnormal Development of Brain and Behaviour.* Baltimore, The Williams & Wilkins Co., 1971, p. 187-203.

Application of a standardized neurological test and analysis, from video-audio tapes, of "free-field" motor, visual, and verbal behavior of 150 "clinically normal" preschool children observed in varied laboratory settings—in order to determine effects of pre- and peri-natal complications on later development—showed that children could be grouped according to "optimal" neurological signs. Neurological condition and behavioral aspects—especially in boys—were significantly related, with nonoptimal boys showing more motor-activity fluctuation in response to absence of sensory input than optimal boys. In novel situations, optimal girls revealed more support-seeking behavior with mother than did nonoptimal girls. A definite decrease in visual exploration over a period of time in an empty room was noted in the nonoptimal boys and girls. In analysis of children's behavior in relation to neurological findings, Ss' sex must be taken into account. (11 refs.) - *B. Berman.*

2167 **FREEDMAN, D. G.** Genetic influences on development of behavior. In: Stoelinga, G.B.A.; & Van Der Werff Ten Bosch, J. J., eds. *Normal and Abnormal Development of Brain and Behaviour.* Baltimore, The Williams & Wilkins Co., 1971, p. 208-229.

Although different cultural practices differentially reward babies' behavior, it is also probable that genetic predispositions affect the cultural norms. Precise genetic studies of behavior, however, are not yet feasible; we can only impute genetic causes to differences found in experimental groups. Investigations of infant-twins have shown individual genetic variation in social behavior, as have studies with the Infant Behavior Profile (the Social Scale shows differences in smiling, vocalizing, and attending to mother), and research in fear of strangers. Sex differences, especially in social modality, are a logical extension of behavior genetics, reflecting the influence of XY and XX chromosomal makeup. Genetic variation within any group accounts for much of the individual variation seen, and gene-pool differences between groups account for group differences. Thus, a series of behavior studies of newborns of various ethnic groups—using 28 general behavior items—has shown differences between orientals and European-American infants in various emotional, temperamental, and intellectual traits. Studies of Navajo and Pueblo infants have confirmed these findings. (25 refs.) - *B. Berman.*

2168 **RYAN, JOANNA.** Language research in the severely subnormal. *Teaching and Training,* 9(1):5-8, 1971.

To explore possible reasons behind the gap in language ability between SMR children and nonretarded children of the same MA, language development was studied in 15 mongol and 15 nonmongol SMRs. CAs ranged from 5 to 9 years, MAs 2.5 to 3.5 years, and IQs from 30 to 49. A control group of normal children had CAs ranging from 2.5 to 3.5 years and IQs of 85 to 125. All groups were matched by sex distribution and socioeconomic class. SMRs with totally unintelligible speech were excluded. Each child's spontaneous speech while playing with various toys was tape-recorded in 3 separate half-hour sessions. Picture vocabulary for everyday objects was assessed in both a naming and a pointing test. Understanding of grammatical distinctions was determined by presenting sets of 2 pictures which differed only in 1 essential respect and asking the Ss to point to the one described by the experimenter. Some memory tests were also included. Although the results have not been fully analyzed, preliminary findings show that the children varied greatly in the amount they spoke, but that each child was fairly consistent across the 3 sessions; there were no differences between groups. Mongol children tended to speak in much shorter sentences than nonmongol ones, who used shorter sentences than the normal children. There was great variation within the nonmongol group, however, some coming within the range of the control group. With the differences in length of sentences, there goes a corresponding difference in the level of grammatical complexity. The 3 groups show no difference in picture vocabulary scores, but when it comes to understanding more abstract kinds of single words, e.g., prepositions, the SMRs do worse than the normals. Thus, while the 3 groups were equal in their knowledge of single words for objects, they differed greatly in their ability to string words together into sentences. It is conceivable that learning the meaning of single words is much easier than combining them meaningfully, so that SMRs because of their limited ability tend to learn many more single words before they eventually combine them than do normal children. (No refs.) - *J. C. Moody.*

University of Cambridge
Cambridge, England

2169 MATTINSON, JANET. *Marriage and Mental Handicap.* Pittsburgh, Pennsylvania, University of Pittsburgh Press, 1970, 231 p. $6.00.

A detailed study of 32 marriages between MR persons in England, conducted by a senior caseworker at the Institute for Marital Studies in London, has attempted to determine the potential viability of such marriages and the degree of their expected dependence on social services. All parties involved had at one time or another been patients at the same mental hospital. Excerpts from taped interviews with the Ss describe in their own words personal feelings about their handicap and its treatment, and their concern for themselves, their spouses, and children. Additional background information, descriptions of methodology, details on the present social situation of the couples, and a general examination of the interpersonal relationships characteristic of these marriages are also included. Conclusive findings as to why some had succeeded and others failed were impossible to formulate because of the small sample size and the absence of usable statistical information. Neither was it possible to psychometrically assess the offspring of these marriages, as had been originally planned. - *N. Mize.*

2170 TALKINGTON, LARRY W.; HALL, SYLVIA M.; & CLELAND, CHARLES C. Behavioral correlates of noncommunicators. *Psychological Record,* 21(2):213-217, 1971.

Behavior characteristics of noncommunicating (NC) MRs were clearly antisocial in comparison with those of a group of communicating (C) peers. A group of NC MRs was compared with a matched group of C peers in 10 behavior categories. The NC group rated higher on variables of aggressiveness and preferred solitude, while the C group achieved significantly higher ratings on variables of interaction, participation, and feeding and dressing skills. A frustration aggression interpretation was discussed, along with the need for intervention techniques to instill communication skills. (31 refs.) - *S. Glinsky.*

Fairview Hospital and Training Center
Salem, Oregon 97310

2171 LENNEBERG, ERIC H. The natural history of language. In: Eliot, J., *Human Development and Cognitive Processes,* New York, Holt, Rinehart and Winston, 1971, p. 296-323.

Man seems to have a biological and evolutionary predisposition to language, and the onset of language may reasonably be attributed to a maturational process. Speech and language exhibit a certain regularity of onset related to age, environment, utility, and degree of practice. Evidence based largely on clinical experience with acquired aphasia indicates that the primary acquisition of language is dependent upon a certain developmental stage which is quickly outgrown at puberty. Studies of mongoloids suggest that even in the absence of structural lesions of the brain, progress in language learning comes to a standstill after maturity. The study of acquired deafness during childhood and later life gives further insight into the importance of age in language acquisition. Absolute values, such as brain weight, are irrelevant to the capacity for language. The hypothesis is advanced that the capacity for language acquisition is intimately related to man's peculiar maturational history and the unique degree of lateralization of function. (No refs.) - *S. Glinsky.*

No address

DEVELOPMENTAL ASPECTS – Psychodiagnostics

2172 ANGENENT, H.L.W. A frame of reference in the observation of institutionalized subnormals. *Journal of Mental Subnormality,* 16(31):107-112, 1970.

The dimensions of a common frame of reference used by the staff of an institution for the MR in evaluating their children were: intelligence, vital and egoistic needs, agreeableness, and excitability.

One hundred institutionalized MRs (39 females; 61 males; CA range 8 to 21 yrs) were individually rated by at least 3 staff members for the presence or absence of 67 behavioral characteristics. Basic dimensions were identified by factor analyzing the correlations between items. (8 refs.) - *J. K. Wyatt.*

No address

2173 **MALONEY, MICHAEL P.; & WARD, MICHAEL P.** Bender-Gestalt test performance of "organic" and "functional" mentally retarded. *Perceptual and Motor Skills*, 31(3):860, 1970.

The performance of functional MRs on the Bender-Gestalt test was significantly ($p<.05$) better than that of organic MRs. Ss were 18 MRs diagnosed as organic (mean CA 13.22 yrs; mean IQ 44.11) and 16 MRs diagnosed as functional (mean CA 14.06 yrs; mean IQ 47.94). A modification of the Koppitz scoring system was used. Functional MRs had a mean score of 22.19, while organic MRs had a mean score of 14.89. These data suggest that the Bender-Gestalt may be a useful ancillary method of identifying organic etiology in MRs. (2 refs.) - *J. K. Wyatt.*

No address

2174 **WOO-SAM, JAMES.** Note on Wechsler Comprehension and Picture Arrangement scores of head-injured epileptics. *Perceptual and Motor Skills*, 31(3):818, 1970.

Scores on the Picture Arrangement subtest of the Wechsler-Bellevue Full Scale tended to be above average for head-injured epileptics, while Comprehension scores tended to be below average. Ss were 23 head-injured veterans (CA range 27-34 yrs; IQ range 89-115; yrs of education 12-14). Epilepsy was established by detailed medical examination, case history report, and EEG findings in 9 Ss. These findings do not substantiate those of previous studies. However, earlier research did not differentiate between types of epileptics or investigate homogeneous groups with regard to age, intelligence, or education. (4 refs.) - *J. K. Wyatt.*

Rancho Los Amigos Hospital
Downey, California

2175 **INGRAM, ROBERT A.** Brain dysfunctional adults. *Medical World News*, 11(40):12, 1970. (Letter)

Children with minimal brain dysfunction grow up to be adults with persisting brain damage which requires continuing medical management, including the administering of anticonvulsants or medication directly related to the dysfunction pattern. The test of choice in identifying these individuals is the Minnesota Multiphasic Personality Inventory, which shows a remarkably high correlation with neurologic EEG abnormalities. (No refs.) - *B. Berman.*

No address

2176 **GROSS, JERRY C.; JOINER, LEE M.; HOLT, LAURENCE E.; & SOMMERS, PAUL.** A kinesio-perceptual test's reliability and validity with retarded subjects. *Journal of Special Education*, 4(2):223-231, 1970.

Items on a kinesio-perceptual test that demanded association and integration by MR Ss had less stability over time than items that measured 'pure' neuromuscular functions. A kinesio-perceptual abilities test (KPT) was administered to 120 institutionalized MRs (CA range 8-20 yrs) and to 105 public school special class Ss (CA range 7-19 yrs; IQ range 40-85). The Wide Range Achievement test was administered to public school Ss. A random sample of 31 Ss was retested 1 month after initial testing. Inter-observer coefficients on the 25 KPT items ranged from .68 to .99. The range for stability coefficients was .2990 to .9911. Of 12 coefficients, 7 were $>.70$. The findings of a cross validation study questioned the validity of using kinesio-perceptual tests to predict academic achievement. (13 refs.) - *J. K. Wyatt.*

Minneapolis Public Schools
Minneapolis, Minnesota

2177 **GARDNER, LESLIE.** Assessment and outcome. *Special Education*, 59(4):11-14, 1970.

A social worker, a pediatrician, and a psychologist on an assessment panel summarized the

educational prospects of 379 cerebral palsied children (CA range 4-11 yrs) with reasonable accuracy. Psychometric scales used included the Stanford-Binet, the Wechsler Intelligence Scale for Children, the Merrill-Palmer Scale, the Columbia Mental Maturity Scale, Raven's Matrices, and the Peabody Picture Vocabulary Test. Accuracy of prediction from these assessments was measured by the type of schooling or training the child was receiving 5 years later. Of 203 children available for follow-up and for whom a fairly definite prediction was made, 81% were receiving the recommended type of schooling. Agreement rates were highest for children recommended for schools for the cerebral palsied at a normal intelligence (82.5%), for children recommended for schools for the cerebral palsied at an EMR level (87%), and for children recommended for placement in a school at the SMR level (88.5%). Predictability was lowest for non-cerebral palsied children recommended for schooling at the EMR level (58%). Factors which may have affected agreement rates included unavailable schooling, family difficulties, assessment and interviewing errors, and changes in the child. (10 refs.) - *J. K. Wyatt.*

No address

2178 GOODSTEIN, HENRY A. Performance of mentally handicapped and average-IQ children on two modified cloze tasks for oral language. *American Journal of Mental Deficiency*, 75(3):290-297, 1970.

A quantitative deficit in cloze performances of low IQ children indicated organizational inefficiency in grammatical decoding processes. The oral language behaviors of public school EMRs and average IQ children were compared by means of 2 modifications of cloze procedure, a production and a recognition task. The Ss were 56 EMRs and 49 average-IQ children who were matched for MA at CA 9 years, 5 months and reading age at CA 8 years, 6 months. The results indicated that EMRs were significantly poorer than average-IQ children in cloze performance, that the recognition cloze task was significantly easier than the production cloze task, and that no qualitative differences were found in the performance of the 2 groups. This reflects inefficiencies in the organization and retrieval of linguistic stimuli of low-IQ children. The investigation pointed up the need for standardized, reliable instruments for assessing various aspects of oral language behavior and the need to transform these findings into effective experimental training programs. (15 refs.) - *S. Glinsky.*

University of Connecticut
Storrs, Connecticut 06268

2179 ACKERMAN, PEGGY T.; PETERS, JOHN E.; & DYKMAN, ROSCOE A. Children with specific learning disabilities: WISC profiles. *Journal of Learning Disabilities*, 4(3):150-166, 1971.

Children with learning disabilities had reliably lower scores than a normal control group on the Arithmetic, Digit Span, Information, and Similarities subtests of the Wechsler Intelligence Scale for Children (WISC). Ss were 82 boys and girls with learning disabilities (CA range 8 to 11 yrs; mean IQ 104) and 34 academically adequate boys (CA range 8 to 11 yrs; mean IQ 111). Experimental group Ss had failed a grade in school; faced impending failure; had reading, spelling, or arithmetic scores 1 year below grade level; or faced impending failure because of severe dysgraphia. All Ss were given the WISC, the Gray Oral Reading Test, the Bender Visual-Motor Gestalt Test, and a special integrative neurological examination. Activity level was determined, and all Ss executed a series of visual-motor conditioning and discrimination tasks. Ss with learning disabilities had significantly lower Verbal Scale scores and higher Performance Scale scores on the WISC than did control group Ss. Serious reading difficulty was related to verbal deficiencies on the WISC, although children with a Verbal or Performance IQ in the low average or dull normal range generally had academic difficulties. Quotients on the Gray Oral Reading Test were significantly correlated with Verbal IQ, Full Scale IQ, Information, Arithmetic, Similarities, Digit Span, and Coding. Neurological status and activity level were not related to WISC or Gray Oral Reading Test scores. (21 refs.) - *J. K. Wyatt.*

University of Arkansas Medical Center
Little Rock, Arkansas 72201

2180 PALKES, HELEN; & STEWART, MARK. Intellectual ability and performance of hyperactive children. *American Journal of Orthopsychiatry*, 42(1):35-39, 1971.

Intelligence, school achievement, and perceptual-motor performance were studied in 32 children with the hyperactive child syndrome (behaviorally defined) and 34 matched normal controls. The hypersensitive children had significantly lower ($p<.001$) mean Verbal, Performance, and Full-Scale IQs on the WISC, but also showed significantly more variation than the control group on the WISC Performance IQ and the WISC Full-Scale IQ. Scores on school achievement tests (Wide Range Reading Achievement Test and Gray Oral Reading Test) and perceptual-motor tests (Bender-Gestalt Test, Developmental Score, and Hidden Figures Test) did not differ from those of controls when adjusted from WISC Full-Scale IQ. Results suggest that hyperactive children have an intellectual as well as behavioral handicap, but that they do learn schoolwork at the rate normal for their level of intelligence. The findings also throw doubt on the idea that many hyperactive children with specific learning disabilities have perceptual-motor handicaps. (11 refs.) - *J. C. Moody.*

St. Louis Children's Hospital
St. Louis, Missouri 63110

2181 **THOMAS, ALEXANDER; HERTZIG, MARGARET E.; DRYMAN, IRVING; & FERNANDEZ, PAULINA.** Examiner effect in IQ testing of Puerto Rican working-class children. *American Journal of Orthopsychiatry*, 41(5):809-821, 1971.

The WISC scores of 116 school age (CA 6 to 16 years) working-class Puerto Rican children in New York City were markedly affected by differences in style between two examiners who were equivalent as to sex, ethnicity, fluency in Spanish and English, and clinical experience. Higher performance level occurred with warm, friendly examiner behavior that encouraged active participation, verbalization, and repeated effort on the child's part as compared to the other examiner's neutral emotional relationship with the children, involvement limited to the test material proper, and no encouragement to try whenever "I don't know" was the first response. IQ scores obtained by the latter examiner correlated better with reading achievement as measured by the Metropolitan Reading Achievement tests, however, probably because little or no opportunity for maximizing performance is possible during the administration of such group tests. It is concluded that the performance level of disadvantaged children on standardized tests of intellectual functioning can be raised by employing examination procedures that are congruent with their spontaneous cognitive styles. This suggests that the academic achievement of such children may also be improved by the use of similar techniques of instruction in school. (17 refs.) - *J. C. Moody.*

New York University Medical Center
New York, New York 10016

2182 **KAPPELMAN, MURRAY M.; LUCK, ELIZABETH; & GANTER, ROBERT L.** Profile of the disadvantaged child with learning disorders. *American Journal of Diseases of Children*, 121(5):371-379, 1971.

A profile of disadvantaged children enrolled in a comprehensive diagnostic and remedial learning disability clinic confirms earlier findings that substantial percentages of these children have an organic, neurologic basis for their educational problems. An analysis was made of the first 100 disadvantaged children referred to a community multidisciplinary Learning Disability Clinic. In this population 33% had organic perceptual handicaps with a preponderance of visual-motor difficulties and 35% had functional area psychosocial disorders. Secondary diagnoses of overlay psychosocial disorders were made in an additional 34% of the population studied necessitating intervention. Boy:girl ratio was over 2:1 in the overall group, 3:1 in the psychosocial group, and equal among the MRs. Eighteen percent of the children had a potentially significant medical history, and 4% a potentially significant prenatal history. This underscores the need for extensive and thorough history-taking. Such a learning disability clinic can only succeed to the extent that remediation areas exist to carry out its recommendations. (5 refs.) - *S. Glinsky.*

University of Maryland Hospital
Baltimore, Maryland 21201

2183 **EDWARDS, R. PHILIP; ALLEY, GORDON R.; & SNIDER, BILL.** Academic achievement and minimal brain dysfunction in mentally retarded children. *Exceptional Children*, 37(7):539-540, 1971.

MR children with minimal brain dysfunction (MBD) are impaired only in certain performance areas as a result of MBD. The performance of MBD children on standardized tests of academic achievement for MR children was investigated. The Ss were 68 children with an IQ of from 55 to 84 and an MA of between 5.0 and 10.11 years, who had diagnosed visual-motor or neurological impairment. The Ss were classified into 3 groups related to type of impairment and a fourth control group with no impairment and were given the WRAT subtests on word recognition, arithmetic, and spelling. Three analyses were performed based on different groupings of the children. The results of the analyses suggest that the degree of academic achievement of MR children in arithmetic and spelling is related to visual-motor impairment, that signs of MBD are not directly related to MRs' reading achievement, and that academic achievement in MRs may become apparent only when test scores are controlled for MA. (3 refs.) - *S. Glinsky.*

Western Carolina University
Cullowhee, North Carolina

2184 **FELDMANN, HARRY.** Psychomotor assessment and rehabilitation of socioculturally deprived children. *Acta Paedopsychiatrica,* 37(9/10):268-293, 1970.

Given the fact that certain sociocultural influences, in the absence of physical or mental disturbances, may prevent a normal cultural adaptation, the testing of children from culturally deprived circumstances by means of static tests based on examination of mental level or intellectual efficiency is inadequate. The correct measurement of development in such children can be made only by means of a dynamic examination of the learning capacity. Educability and the capacity for learning must be examined as a function of psychomotor potential and of plasticity of the neurological substratum. The use of the method of psychomotor education based on Jaques-Dalcroze rhythmics (in which the influence of music is considerable) has greatly facilitated the successful acquisition of motricity, which in turn enables the child to better control intellectual development and socialization in a new environment. Three case histories of work with culturally-deprived MR subjects illustrate the usefulness of the 48 progressive exercises based on imitation, personal investigation, and mutual effort which comprise the rhythmics method. Only through these means is it possible to judge the evolutive potential of the child and to make a dynamic diagnosis of his future educability. (33 refs.) - *P. Custers.*

6 Cours de Rive, CH-1204
Geneva, Switzerland

2185 **BUDOFF, M.** Learning potential: Assessing ability to reason in the educable mentally retarded. *Acta Paedopsychiatrica,* 37(9/10):293-310, 1970.

A large proportion of those labeled MR from a low income background are academic risks in a traditional school setting. But this failure to learn via traditional methods cannot simply be extrapolated to indicate a generalized inability in adult life if these children are permitted to bypass their deficient literary skills in the occupation they choose. From this analysis is derived the concept of educable mental retardation (EMR), a largely poverty and school-related condition that has a low incidence prior to the start of school or following school leaving. The AAMD classification which distinguishes separate parameters of measured intelligence and adaptive behavior does not adequately make this distinction because adequacy of adaptive behavior in school age children is defined as the adequacy of their behavior in school. The learning potential assessment procedure recommended here was devised as an attempt to avoid this circularity. The question asked is whether these children can reason adequately regardless of school performance. To minimize the negative effects of differences in prior experiences, the students are taught principles relevant to nonverbal reasoning problems, following a pretest trial. Test results range from children with high scores on both tests to those who perform poorly on the pretest and do not improve following training. This last category (non-gainers) is considered to represent true MR. Research into the more general validity of this testing procedure is continuing. (12 refs.) - *P. Custers.*

Research Institute for Educational Problems
Cambridge, Massachusetts 02139

2186 **FREEDMAN, D. G.** Behavioral assessment in infancy. In: Stoelinga, G.B.A.; & Van Der Werff Ten Bosch, J. J., eds. *Normal and Abnormal Development of Brain and*

Behaviour. Baltimore, The Williams & Wilkins Co., 1971, p. 92-99.

Infant tests generally have poor predictive value for later development. The widely used Gesell Test yields only a .50 reliability predictive value up to 5 years of age. Reasons for such poor results are that motor items in infancy usually involve the whole body, which is less true in later years; "mental" items in infancy are chiefly assessments of eye-hand coordination and overall sensory alertness; standardized scales cannot evaluate developing personality traits; and infant tests are chiefly survey instruments. More sensitive tests might improve prediction. New insights into infant capacity are now deriving from outside the testing movement. Work in hypothesis testing and theory development is producing new measurement devices, which are stimulating increased study of infant behavior. (9 refs.) - *B. Berman.*

2187 **BRAZELTON, T. B.; & FREEDMAN, D. G.** Manual to accompany Cambridge Newborn Behavioral and Neurological Scales. In: Stoelinga, G.B.A.; & Van Der Werff Ten Bosch, J. J., eds. *Normal and Abnormal Development of Brain and Behaviour.* Baltimore, The Williams & Wilkins Co., 1971, p. 104-132.

A manual to accompany the Cambridge Newborn Behavioral and Neurological Scales—a test to distinguish differences among normal newborns—stresses that the infant be examined under relaxed conditions (in the lap with head well up, ideally between feedings). State of consciousness (various sleep and awake states) is one of the most important variables in the observation period. The items measured by the test on point scales include: general tonus, skin-color lability, peak of excitement, lability of states, alertness, following inanimate object with head and eyes, reaction to sound, defensive movement, irritability, self-quieting activity, consolability with social intervention, pull to sit, head movement in prone, activity, social interest, smiling, leg and arm movements, habituation to stimulation, hand and mouth facility, tremulousness, startle, vigor, cuddliness, and a variety of elicited reflex movements. (5 refs.) - *B. Berman.*

2188 **HENNIG, HEINZ.** Zur psychodiagnostischen relevanz des abzeichentests und des baumtests bei der differenzierung der schwachsinnsgrade im kindesalter (The psychodiagnostic relevance of the copy drawing test and the "tree drawing test" for the differentiation of the degree of imbecility in children). *Psychiatrie, Neurologie, und Medizinische Psychologie,* 23(2):94-100, 1971.

The severity of childhood MR can be estimated by performance on tests of copying a drawing and drawing a tree. Copying a series of geometric figures of increasing complexity was impossible for imbeciles; the mildly MR differed from normal only on the most complex figures. In drawing trees, the MR children all produced "early" forms as previously described by Koch. (No refs.) - *E. L. Rowan.*

Martin Luther Universitat Halle
402 Halle, Germany

2189 **PAULUS, DIETER H.; & RENZULLI, JOSEPH S.** A factor-analytic study of the First Grade Screening Test. *Psychology in the Schools,* 8(1):57-59, 1971.

The First Grade Screening Test (FGST) samples handicaps that might impede school success. Educational and psychological development are evaluated in the areas of accumulated fund of knowledge, body image, perception of emotional maturation, accuracy of perception of parental figures, perception of appropriate play, visual-motor coordination, ability to follow directions, and memory. An empirical and rational investigation of the factor structure of individual items on the FGST examines their relationship to these areas. The factor structure of the FGST based on rational and theoretical data is compared with the factor structure based on empirical relationships discovered in the data. (3 refs.) - *J. K. Wyatt.*

No address

2190 **LEVINE, EDNA S.** Mental assessment of the deaf child. *Volta Review,* 73(2): 80-105, 971.

The examiner of intelligence in deaf children must be particularly sensitive to the background

needs and capabilities of the pupils to be tested and carefully consider certain guidelines in the selection and administration of tests. The fixed IQ tests, especially for the deaf, do remain a valid tool, if used with great caution. Although additional psychological and intelligence tests for the deaf do exist now, there were only 11 tests for the deaf out of over 2,000 for the hearing in 1961. Clinical controls or considerations, which can make even a questionable test for the hearing a more effective assessment device for the deaf, include securing the collaboration of other examiners experienced in testing the deaf, careful study of scores, especially for items not revealed by numerical scores, and reporting IQ scores for recording only when they are within a valid range of estimate, while conversely providing a personal estimate when test scores are not submitted. Additionally, one should arrange for retests, secure evaluative data from other sources, and follow up on recommendations derived from test results. These measures will help the examiner obtain the best possible results from the use of standardized tests on the relatively unstandardized population of the deaf. (36 refs.) - *S. Glinsky.*

No address

2191 **WALTI, ULRICH.** Der Entwicklungstest nach O. Brunet und I. Lezine als Untersuchungsmethode zur Beurteilung des psychomotorisch retardierten Kleinkindes (The development test by O. Brunet and I. Lezine as an investigation method to evaluate young children with psychomotor retardation). *Schweizerische Zeitschrift fur Psychologie und Ihre Anwendungen*, 30(1):21-39, 1971.

Evaluations of young children with psychomotor retardation by means of the Brunet-Lezine development test failed to identify several distinct pathological groups. The Brunet-Lezine development test was given to 104 motor retarded and 30 normal infants. The retardation of the motor retarded was strongest in the subscale of language development, whereas the subscale of gross motor development was relatively less affected. The Brunet-Lezine test, constructed to evaluate mental development in early ages according to the 4 factors defined by Gesell, failed to differentiate between several clinically defined pathological groups. Especially in view of therapeutic decisions, the use of more sensitive methods of psychological diagnosis in this area seems desirable. (24 refs.) - *S. Glinsky.*

Abteilung fur Entwicklungsstorungen
CH-3000 Bern, Switzerland

TREATMENT AND TRAINING ASPECTS – Educational

2192 WILLIAMS, MAIR. How do I teach art? *Parent's Voice*, 20(4):24-25, 1970.

Among the techniques used in art education with MR children are observation lessons, color recognition, scribbling, texture identification, and the use of basic shapes. Group activity can be used for friezes or stage backdrops. The materials should be geared to the level of the children. (No refs.) - *J. K. Wyatt.*

No address

2193 ASHTON, A. W. 'Transfer': What needs to be done? *Parent's Voice*, 20(4):6-7, 1970.

Educational programs for MRs should educate every child to his fullest capacity, discover the possible horizons for each individual, and identify the best teaching techniques for each child. Adequate numbers of well trained teachers are needed, as well as the services of ancillary staff and consultants. Specialized buildings designed to meet the specific needs of MRs should be provided in sufficient numbers. Educational programs should aim at changing community attitudes toward the MR so that these communities will assume responsibility for MR children and provide adequate programs for them. (No refs.) - *J. K. Wyatt.*

Assistant Education Officer
Berkshire, England

2194 BEASLEY, NIGEL; & HEGARTY, JOHN. The use of the Touch-Tutor with SSN children. *Journal of Mental Subnormality*, 16(3):113-118, 1970.

Two training sessions did not increase the number of SMR children able to match-to-sample in a machine teaching program employing the Touch-Tutor. Under the "Sound" condition, correct responses were followed by the naming of the correctly matched picture, and the machine advanced 5 seconds after either a correct or incorrect response. In the "No Sound" condition, the slide advanced immediately after a correct response, but the machine made no verbal response. One-choice, 2-choice, and 3-choice slides were presented in either a progressive or mixed sequence. The training sessions were taught under "Sound Progressive," "Sound Mixed," "No Sound Progressive," or "No Sound Mixed" conditions. Ss were 35 SMRs (CA range 8 to 19 yrs). No one teaching condition proved more rewarding or attractive, and pre- and post-test performance generally did not improve. In some Ss the machine seemed to reinforce a variety of bad habits in responding. (9 refs.) - *J. K. Wyatt.*

No address

2195 MARTIN, MADELYN H. The slow learner: Building the slow learner's self-esteem. *Today's Education*, 59(3):46-47, 1970.

The slow learner's self-esteem must be developed through concerted efforts on the teacher's part to praise strengths and positive aspects of behavior, and avoid criticism, ridicule, and negative reactions. Examples of the ways in which an elementary school can improve the self-esteem of the slow learner are cited. (No refs.) - *D. Fergerson.*

A. M. Elementary School
Richmond, Virginia

2196 KARNES, MERLE B. The slow learner... What are his characteristics and needs? *Today's Education*, 59(3):42-44, 1970.

The slow learner – sometimes called dull-normal or intellectually backward – falls between the

normal and the educable MR in learning speed (IQ 75-90). He differs from the average in having more physical defects, poorer reasoning ability, shorter attention span and less retention, and less developed language and communication skills. He is consistently below academic grade level, showing inadequate work habits and motivation, difficulty in following directions, and absence of curiosity and creativity. The slow learner generally comes from a disadvantaged home and will display lack of self-confidence and minimal, if any, leadership qualities. (No refs.) - *B. Berman.*

University of Illinois
Urbana, Illinois 61801

2197 FOY, BOB. The slow learner: Reaching the unreached. *Today's Education,* 59(3):44-46, 1970.

Education of slow learners can be successful if the curriculum is geared to their specific academic difficulties and if realistic vocational training is provided. It is necessary to accept the limitations (such as poor reading level of the teenage slow learners) and concentrate on developing other areas of competence. Slow learners need school-sponsored work-experience programs in which they can earn money while learning and also increase their self-confidence. (No refs.) - *D. Fergerson.*

Pacific Prevocational School
Seattle, Washington

2198 Education for autistic children. *Lancet,* 2(7683):1147, 1970.

At least 100 educational units for autistic children, evenly distributed throughout the country, are needed in Great Britain. Of the 32 present units, 22 are in southeastern England; 13 are in the London area. There are no units in Scotland or Wales. There are an estimated 3,772 autistic school-age children and 2,155 of less than school age in Britain. There are about 2,000 autistic teenagers, yet there is only one unit for this population. Autistic children should have special schooling but live at home. This cannot always be achieved with the present lack of facilities. (1 ref.) - *E. Kravitz.*

2199 APPLEGATE, DOROTHY. Ten-point reading program for educables. *Instructor,* 79(2):140-141, 1969.

In a reading program for educable retardates, one must allow for their limited conceptual experiences and make the reading experience pleasant by ensuring small "successes." Special education teachers should follow these procedures: (1) do not group pupils, (2) do not teach all reading skills simultaneously, (3) activate pupil thought processes, (4) give equal emphasis to all learning phases, (5) encourage children to ask for help, (6) plan creative activities, (7) keep walls free of a mass of self-helps, (8) do not put words into a child's mouth, (9) look out for loafers, and (10) make no comparisons among pupils. (No refs.) - *B. Berman.*

Jefferson School
Sharon, Pennsylvania

2200 JOHNSON, K. E. Increasing the capacity to learn. *Slow Learning Child,* 17(3):163-169, 1970.

The premise that retardates are qualitatively different from normals is generally accepted; however, if it is assumed instead that their deviation is quantitative – one of degree – then, if this degree can be measured, the elements that make up intellectual potential can be elevated and the learning capacity of retarded children can thus be improved. Such an assessment, which is more relevant to educational processes than is IQ, suggests the possibility of developing learning capacity through educational processes rather than by training in cases of moderate retardation. Reading then becomes a form of contact with surroundings and a meaningful source of pleasure – not just a survival technique. Two examples of detailed assessment and development are cited, in which improvement in the quantitative status of deficient visual perception brought improved reading capability. (No refs.) - *B. Berman.*

Cromehurst Special School
New South Wales, Australia

2201 EDWARDS, JOSEPH; & *STERN, CAROLYN. A comparison of three intervention programs with disadvantaged preschool children. *Journal of Special Education,* 4(2):205-214, 1970.

The effects of a modified version of the UCLA Preschool Language Program, the Readiness for Language Arts program (BRL), a structured placebo program, and a regular control group with preschool disadvantaged children were compared. Ss were 74 girls (mean age 51.7 mos; mean IQ 75.3) and 89 boys (mean age 51.8 mos; mean IQ 75.5) who were enrolled in Head Start programs. Pre- and post-test data were available on 104 Ss. On the Preschool Inventory, experimental group Ss performed significantly better than control group Ss, and the performance of UCLA-group Ss was superior to that of BRL- or placebo-group Ss. BRL-group Ss had significantly superior scores on the Visual Discrimination Inventory. The UCLA group had significantly higher scores than the placebo group on 2 subtests of the UCLA tests (Picture Reading and Home Environment), and the UCLA and BRL groups had significantly higher scores than the placebo group on the Problem Solving subtest. Anglo-American children had significantly higher scores than Afro-American or Mexican-American Ss on the Peabody Picture Vocabulary Test. On the Visual Discrimination Inventory, the scores of Mexican-American Ss were significantly higher than those of Afro-American Ss. Partial support was obtained for the hypothesis that the Preschool Language Program would bring gains in a wider range of cognitive skills. An unanticipated finding was the significant superiority of the BRL group on the Visual Discrimination Inventory. Teachers with the greatest amount of preschool teaching experience produced generally superior results. (31 refs.) - *J. K. Wyatt.*

*University of California
Los Angeles, California

2202 **ELLIOTT, RAYMOND N., JR.** Meaningfulness in school tasks for EMR children. *Journal of Special Education*, 4(2):189-197, 1970.

Meaningfulness of materials facilitates learning in normal and MR Ss. Learning tasks based on meaningful material were presented to 47 special class EMRs (mean CA 9.4 yrs; mean IQ 63.61; mean MA 6.4 yrs), 85 normal children matched for CA, and 89 normal children matched for MA. Ss involved in learning tasks based on nonmeaningful material were 41 special class EMRs (mean CA 9.4 yrs; mean IQ 68.49; mean MA 6.5 yrs), 93 CA-matched normals, and 85 MA-matched normals. Test scores were based on prepresentation data, original learning, and retention. Meaningfulness had a more important influence on the original learning of EMR Ss than on the original learning of normal Ss. The influence of meaningfulness on learning appeared to be different for the 3 groups. MRs evidenced learning and retention scores very similar to those of normal Ss in the meaningful learning situation. Familiar associations or environmental cues can be used to make meaningful material from nonmeaningful material. (27 refs.) - *J. K. Wyatt.*

Pennsylvania State University
University Park, Pennsylvania

2203 **HALL, SYLVIA M.; & TALKINGTON, LARRY W.** Evaluation of a manual approach to programming for deaf retarded. *American Journal of Mental Deficiency*, 75(3):378-380, 1970.

A manual approach on sign comprehension and language development demonstrated significant gains by deaf MRs. The Ss were 30 profoundly hearing-impaired MRs and 30 hearing MRs matched on age, IQ, sex, and length of institutionalization. After pretesting, a 6 month manual approach deaf education program, and posttesting, the deaf group achieved greater gains than the hearing group in both the use of manual sign language and Verbal Language Development Scale performance. The results underline the feasibility of such a manual sign approach, which furnishes both a flexible, simple initial communication system and a system easily assimilated and used by ward personnel and peers. (6 refs.) - *S. Glinsky.*

P.O. Box 1269
Austin, Texas 78767

2204 **PREHM, HERBERT J.; & STINNETT, RAYMOND D.** Effects of learning method on learning stage in retarded and nonretarded adolescents. *American Journal of Mental Deficiency*, 75(3):319-322, 1970.

The learning performance of MRs was inferior to that of nonretarded Ss at each of 3 stages of learning, regardless of method used. The Ss were

60 EMRs with a mean CA of 14.57 years, mean MA of 10.05 years, and mean IQ of 69.05, and also 60 intellectually average children with a mean CA of 13.85 years, mean MA of 14.18 years, and mean IQ of 102.43. Their learning performance was compared as a function of learning stage using a paired-associate learning task. Twenty Ss within each group were randomly given 1 of 3 methods of learning, either the Equal Amount Learned method (EAL), the Modified Method of Adjusted Learning (MMAL), or the Method of Adjusted Learning (MAL). Analysis of variance indicated that the MRs exhibited a deficit at each stage of learning (response learning and associative stages) and that the EAL method prolonged both the response learning stage and the stage of learning wherein the S consistently pairs correct stimulus and response items. Future research should deal with isolation and evaluation of the components of each stage of learning. (11 refs.) - *S. Glinsky.*

University of Oregon
Eugene, Oregon 97403

2205 PENNINGTON, J. A. More work: More rewards. *Teaching and Training,* 8(4):108-112, 1970.

A British educational psychologist looks forward to the challenge of working with SMRs, which has been hitherto partially denied to him. The educational psychologist has until recently been legally prevented from working with SMRs in training centers, since by law the training of SMRs has been the exclusive responsibility of the health and medical authorities. The anticipated passage of the Education Bill in 1971 will transfer all MR education to educational authorities and thus open a most difficult but most rewarding field of endeavor to educational psychologists. (No refs.) - *S. Glinsky.*

No address

2206 Education for the intellectually handicapped. *Intellectually Handicapped Child,* 9(4):17-18, 1970.

Education for the intellectually handicapped should help each child develop his physical, mental and social abilities to their full potential. The Intellectually Handicapped Children's Society has pioneered in such education and has a fully articulated series of educational establishments. These include pre-school centers, occupational groups for situations where 5 or more MRs have no access to schools, short-stay homes, and opportunity workshops for those over 18 years of age. Continuing parental education is vital, as is the development of a program for the training of teachers in special education. (No refs.) - *S. Glinsky.*

2207 CRAGIN, WESLEY E.; & BETER, THAIS R. The mentally retarded child speaks to the movement educator. *Journal of Health Physical Education Recreation,* 42(2):78, 1971.

If the physical, or movement, director were to listen to the retarded child, he would hear the latter's deep call for understanding, for love. Please take my hand, the retardate would say, and lead me into your wonderful land; guide my experiences in learning to move. I am a unified organism—there is much I can do to grow and develop. I need objects to help me understand abstractions; I can develop my sensory modalities. I need physical activities to help me extract from objects information about colors, forms, weight. I need locomotor movements to help me perform everyday activities. You can stimulate my cognitive processes through movement experiences. There is much inside me yearning to be discovered. (1 ref.) - *B. Berman.*

No address

2208 GUESS, DOUG; SMITH, JAMES O.; & ENSMINGER, E. EUGENE. The role of nonprofessional persons in teaching language skills to mentally retarded children. *Exceptional Children,* 37(6):447-453, 1971.

Significantly greater raw score gains on the Illinois Test of Psycholinguistic Abilities were made by 39 MR children (mean CA 12.75 yrs; mean IQ 33; mean MA 3.95 yrs) who attended language training classes over a 2-year period than by a matched control group. Two former psychiatric aides were trained to serve as language developmentalists. Experimental group Ss

were divided into 'high' and 'low' functioning groups. They attended daily one-hour-long classes which included teaching from the Peabody Language Development Kits and a series of lessons designed to meet special deficits and needs. A token reinforcement system was used to shape language responses and control disruptive behavior. Over the first 9 months, language training groups had significantly greater gains in Stanford-Binet IQs ($p<.03$). However, gains over the 2-year period were not significant ($p<.07$). Gains on the Vineland Social Maturity Scale and the Michigan Verbal Language Development Scale were not significant. (10 refs.) - *J. K. Wyatt.*

Kansas Neurological Institute
Topeka, Kansas

2209 **NELSON, CALVIN C.; & SCHMIDT, LEO J.** The question of the efficacy of special classes. *Exceptional Children,* 37(5):381-384, 1970.

In order to cope with the question of the efficacy of special classes, educators must depart from a common sense approach, advance beyond intuitive or *a priori* evaluation, and submit special education constructs to critical examination. Research studies and arguments which seeks to deal with the efficacy question must deal systematically with basic epistemological problems, including the serious consideration of *a priori* questions. Special classes can only be rejected or justified after systematic examination of their effectiveness. The constructs that underlie educational programs should become prior statements that specify the meaning of statements made on the value of special classes. (8 refs.) - *J. K. Wyatt.*

California State College
Fullerton, California

2210 **WILLIAMS, MARGARET L.; & *SCARR, SANDRA.** Effects of short term intervention on performance in low-birth-weight, disadvantaged children. *Pediatrics,* 47(1):289-298, 1971.

A group of 120 preschool children of low birth weight (less than 2.5 kg) and poor socioeconomic status were provided with short-term programs of tutoring with educational materials, the materials alone, or control (no intervention). Neurological impairment was the most significant factor in any gains in motor function, social maturity, or intellectual ability. Tutoring and toys resulted in improved verbal performance in neurologically normal children and improved social maturity in neurologically impaired children. The provision of educational materials alone resulted in no gains. Early and intensive intervention can result in increased achievement in these disabled individuals. (31 refs.) - *E. L. Rowan.*

*University of Pennsylvania
Philadelphia, Pennsylvania 19104

2211 **FRANKS, DAVID J.** Ethnic and social status characteristics of children in EMR and LD classes. *Exceptional Children,* 37(7):537-538, 1971.

An apparently disproportionate number of Afro-American children are in EMR rather than in learning disability (LD) classes. The ethnic and social status characteristics of children in EMR and LD classes, grades one to eight, in the state of Missouri were determined. The Ss were a random sample of 274 EMRs, with a CA range of 6 to 15.4 years and an IQ range of 45 to 93, selected from an EMR population of 2,777, and a sample of 215 LD children, with a CA range of 6 to 14 years and an IQ range of 43 to 129, selected from a state LD population of 360 children. Information was gathered by means of a questionnaire to the special education teacher of each child, requesting the occupation of the principal parental wage earner of each child, the child's ethnic origin, and the child's IQ. The results showed 34.21% Afro-American children and 65.79% white children in EMR classes, while 3.22% Afro-American children and 96.78% white children were in LD classes. These data support earlier findings that EMR classes are becoming repositories for low-income, non-white children, with higher-income, white children to be found predominantly in LD classes. Educators must review EMR and LD class composition and insure equal educational opportunities for low-income, ethnically different children. (6 refs.) - *S. Glinsky.*

University of Missouri
Columbia, Missouri

2212 **FRIEDMAN, RONALD J.; & MacQUEEN, JOHN C.** Psychoeducative considerations of physical handicapping conditions in children. *Exceptional Children*, 37(7):538-539, 1971.

Physical handicap among children is predominantly associated with some degree of MR, which has important implications for the educational programs for the physically handicapped. School age children were evaluated to ascertain the prevalence of psychologically and educationally relevant physical handicapping conditions. Out of 44,943 children, 195 children were identified as physically handicapped, evaluated by a team of specialists in 5 diagnostic clinics, and placed in 5 physical handicap source categories. The results showed 43% with some degree of MR, with 24% in the EMR range and 19% in the TMR range. Hearing examinations revealed a hearing loss percentage equal to that among normal children. In addition 25% displayed some degree of psychological maladjustment, and a significant amount of intra-family difficulty in adjusting to the handicapped child was noted. Educational programs for the physically handicapped child must take into account the MR and emotional disturbances of the child and should work closely with the family. (No refs.) - *S. Glinsky.*

University of Iowa
Iowa City, Iowa

2213 **MacMILLAN, DONALD L.** Special education for the mildly retarded: Servant or savant. *Focus on Exceptional Children*, 2(9):1-11, 1971.

The polarization regarding either complete abolition of EMR classes or complete integration of EMRs into regular classes, based on L. M. Dunn's 1968 article on the subject, is an inadequate response to the complexities of the problem. Recent evidence indicates that although self-contained classes do not achieve the originally expected growth, some low IQ children can still benefit from them. Dunn's method of identifying MR children is strictly a psychometric definition of MR and ignores the consensus AAMD definition. The effects of placement and labelling must not be considered as the sole cause of achievement and adjustment problems, since there is a dearth of evidence for this. Adjustments in the environment which are strictly cognitive in nature are unlikely to aid learning in low IQ children, whose problems are emotional or motivational. Finally, the real issue is the extent to which a wider range of individual difference can be accommodated in the regular class, and not whether special or regular classes are better for EMRs. (38 refs.) - *S. Glinsky.*

University of California, Riverside
Riverside, California

2214 **LANE, PAULINE M.** Individual academic evaluation. *Focus on Exceptional Children*, 2(8):11-14, 1971.

Individualized Performance Profiles in various skills make possible meaningful individual academic evaluation of MRs. Standardized tests, based on normal populations, are inadequate for measuring the small progress increments among MRs. Academic programs for EMRs using the ungraded system of pupil placement function best. The Performance Profile is an adequate instrument for measuring EMR progress in an ungraded situation. It determines the performance goal, based on the skill-level necessary to reach that goal, creates a curriculum to attain the goal, and enables specific evaluation of progress toward the goal. The curriculum content is structured according to a continuum of prerequisite skills and performance levels from basic to most difficult, representing a normal sequence of progress. The ability of the EMR to perform the sequential learning tasks yields his Performance Profile. Such Profiles exist for arithmetic, reading, sewing, homemaking, and shop, and have been highly successful with EMRs. (10 refs.) - *S. Glinsky.*

Arkansas Children's Colony
Conway, Arkansas

2215 **DENO, EVELYN.** Some reflections on the use and interpretation of tests for teachers. *Focus on Exceptional Children*, 2(8):1-11, 1971.

The role and validity of educational tests are being seriously questioned and they must be thoroughly reevaluated regarding their function in the educational process. Present-day opinions concerning such tests call for both less and more

intelligence and personality testing and reliance upon their results. In the past tests have been devised without an adequate theory base vis-a-vis the educational process. In special education children have been grouped according to underlying physical conditions and assigned to classes subsequent to psychometric testing. Present research questions any direct links between physical conditions and learning behavior. Testing must also take into account the effect of environmental variables, including the teacher, upon the child. The teacher as an instructional system is the nexus through which information from various disciplines is amalgamated and transmitted to the child. Emphasis in teaching is shifting to a self-actualization ideal where individuals are judged in comparison to their past performance rather than to a test-derived mean. Accountability demands evidence of children's progress toward goals, but teachers must beware of producing behavior geared primarily toward good test performance. Development of meaningful testing will require a rethinking of the entire educational process, including performance, objectives, behavior, and the ultimate goals of society for the individual. In addition to formal tests, common evaluation of progress by the teacher must not be excluded. (40 refs.) - *S. Glinsky.*

University of Minnesota
Psycho-Educational Center
Minneapolis, Minnesota

2216 **AUERBACH, AARON G.** The poor retarded child. *Deficience Mentale/Mental Retardation,* 21(3):8-10, 1971.

Lack of recruitment of poor retarded children is one of the weaknesses of the nursery schools and day care centers in Canada. Active recruitment of these children should be initiated, and teachers and volunteers must become knowledgeable about the social conditions which affect poor retardates adversely. The development of a general practitioner nursery school teacher depends upon the ability to relate to young children, whether they are normal, disadvantaged, or retarded. To be successful, the teacher may have to deal with community organizations, governmental departments, and curriculum development. Most important, the teacher will be in contact with children he never learned about previously. Through varied experience, the teacher may then be able to select a special field, such as the teaching of disadvantaged or handicapped children. (No refs.) - *B. J. Grylack.*

York University
Toronto, Ontario, Canada

2217 **SHUSTER, SUSAN K.; & SHUSTER, ARNOLD A.** Educational research & development in mental retardation: Problems & directions for Canadians. *Deficience Mentale/Mental Retardation,* 21(3):11-15, 1971.

The primary objectives of Canadian research and development activities in MR should be to develop and investigate modifications of educational practice. Improvements would permit more mildly retarded children to go through school without being identified as retarded and would allow more children who are identified as mildly and moderately retarded during their school years to begin adult life as nonretardates and/or acceptable members of society. To achieve these goals, characteristics of retardates which lead to their labeling must be identified, and factors inherent in the social environment of retardates which deter success must be determined. General research aims should be established to integrate knowledge of current research and development, to initiate new multidisciplinary programs, to utilize results to assist educational practice, and to train personnel skilled in research. (9 refs.) - *B. J. Grylack.*

Indiana University
Bloomington, Indiana

2218 **JUSTICE, WANDA P.** Innovative programs to meet the needs of the pre-school retarded child. *Deficience Mentale/Mental Retardation,* 21(3):4-6, 1971.

Any program concerned with the educational environment of the retarded child in the preschool stage must consider many factors, including the varied needs of the retarded group, the advantages of new methods despite conceptions of children's limitations, and the need to share ideas and goals with parents and professionals from other disciplines. To meet these requirements, a preschool was established to serve 32 functionally retarded children between

the ages of 3 and 6 with conditions ranging from mild to severe retardation due to various causes. Aside from dealing with daily routines, program activities emphasize language, motor, and perceptual development; social skills; emotional growth; self-image and awareness of the environment; and auditory, visual, and tactile discrimination. Workshops are held for parents and professionals from related fields. Short-term gains were realized by the children enrolled in the program. (No refs.) - *B. J. Grylack.*

University of British Columbia
Pre-School Program
Vancouver, British Columbia, Canada

2219 ANDERSON, SUSAN. Current trends in mental retardation films. *Deficience Mentale/Mental Retardation,* 21(3):32-33, 1971.

The first International Film Festival in Mental Retardation, designed as a showcase of the best and most recent North American and European films on MR, was held in conjunction with a recent conference on MR. As more people are preparing to enter the field and as staff retraining or development programs are being introduced, the need has arisen for films dealing in detail with such topics as residential care, workshop management, behavior therapy, social casework, various approaches to special education, physical education, home care, and religious experience. The use of animation and still pictures with accompanying dialogue or commentary constitutes a new trend. The demand for more and better films on MR is increasing, and a regularly scheduled film festival may provide the necessary stimulus. (No refs.) - *B. J. Grylack.*

National Institute on Mental Retardation
Toronto, Ontario, Canada

2220 WHITTAKER, CAMILLA. The institutional library—a teaching tool for retarded children. *Journal for Special Educators of the Mentally Retarded,* 7(2):119-123, 129, 1971.

The librarian at an institution for MR children has responsibility for residential children who do not attend school as well as for those who do. To make the library an effective teaching environment, the level of functioning and personal needs of the children must be evaluated. Following the appraisal, a child can be placed in an appropriate group situation. Books can be used with the preschool child to broaden communicative speech and to aid in the development of an awareness of environment. The child with an MA of 5, 6, or 7 years should be able to recognize everything he sees in the book, and his attention span will be long enough for him to profit from his reading. The older child who cannot read is hindered by an additional problem, for he is defensive on account of his inability. Although the child may never be able to attain a significant measure of reading competence, books can become a source of enjoyment and information for him if his interests are matched with the appropriate material. (No refs.) - *B. J. Grylack.*

The Hissom Memorial Center
Sand Springs, Oklahoma

2221 ABBOTT, ROBERT E. "Junk" material which can be used for training the mental retardate. *Journal for Special Educators of the Mentally Retarded,* 7(2):124-129, 1971.

Awareness of the need for concrete teaching aids in schools for MR children can be attributed to a better understanding of individual differences and of the growth and development process and to increased awareness of the perceptual aspects of learning. Due to the prohibitive cost of some commercial materials, various free materials that are available in almost every household in a school district should be utilized. Such items as egg cartons, old plastic utensils, golf tees, wooden sticks, and rubber jar rings are useful in activities geared towards the teaching and strengthening of visual-motor skills, figure-ground skills in perception, perceptual constancy skills, and spatial relationship skills. It is the responsibility of the creative teacher to use these and similar materials to the benefit of MR children. (No refs.) - *B. J. Grylack.*

Special Education, District #61
Waukegan City School District, Illinois

2222 McCAULEY, JOHN J. Training in analytical listening through arithmetic. *Journal*

for Special Educators of the Mentally Retarded, 7(2):103-105, 1971.

Arithmetical exercises, including speed tests, have been found to provide worthwhile training in analytical listening for EMR teenagers with a mean IQ of 63 in a secondary special education class. The exercises appear to improve concentration, because they demand the immediate selection of a specific course of action. Through careful review of test papers, the teacher can identify areas of difficulty and can bring about significant improvement in the skills of students. It seems to be more difficult to help the same students to make progress in reading ability and comprehension. (No refs.) - *B. J. Grylack.*

Clearcreek High School
Amanda, Ohio

2223 **STIAVELLI, R. E.; & PICKEL, F. W.** The Frostig program for the development of visual perception with two groups of educable mentally retarded children. *Journal for Special Educators of the Mentally Retarded*, 7(2):130-132, 141, 1971.

Fifty-three pupils enrolled in 4 primary level special education classes for EMR students were used in an evaluation of the effectiveness of the Frostig program in improving perceptual skill weaknesses of these students. A test administered to 28 experimental and 25 control children with a mean IQ of 71 was employed to appraise abilities in the areas of eye-motor coordination, figure-ground perception, constancy-of-shape recognition, and discrimination of position in space and spatial relationships. The results of the study indicated that the Frostig program improved eye-motor coordination and form-constancy discrimination significantly in the experimental group. Although the program was not as effective as predicted, the teachers of the experimental classes reported noticeable improvement in student performance on visual motor and perceptual tasks and in academic areas as well. The Frostig materials and techniques are valuable for specific, identified learning problems, as many remedial approaches are, and should not be used with all students to remediate all learning difficulties. (2 refs.) - *B. J. Grylack.*

Sacramento City Unified School District
Sacramento, California

2224 **TREMBLEY, PHILLIP W.** Changing concept of intelligence and its effects on future class organization. *Journal for Special Educators of the Mentally Retarded*, 7(2):74-78, 89, 1971.

Limitations imposed by traditional categorization of children with learning problems have led the Educational Services Center to devise a curriculum for these children based upon grouping by shared deficiency areas. Special education will be involved in deficit removal so that each child can fulfill his potential. Classes will not be arranged according to IQ. The main underlying philosophy of the new curriculum rejects the premise that the concept of MR is meaningful for the solution of individual learning problems and that it is a fixed condition which is not remediable. Instead, a curriculum emphasizing that intelligence is the function of assets and deficits and that deficits are definitely remediable will be instituted. (No refs.) - *B. J. Grylack.*

Educational Services Center
Granville, New York

2225 Tips for teachers. *Journal for Special Educators of the Mentally Retarded*, 7(2):79-81, 1971.

Music and dance are essential for the wholesome development of MR children. TMR and EMR children derive particular pleasure from using rhythm instruments. The special class teacher should take advantage of parties and festivals and should sing, dance, and play instruments together with the children and use the record player and radio in the classroom. As an extremely effective device for encouraging oral communication, storytelling is an essential skill for socialization and should be a regular part of the school program. Humor, action, illustration, and audience participation are important ingredients of storytelling which will stimulate interest. (No refs.) - *B. J. Grylack.*

2226 **VIGGIANI, JAMES C.** Rochester's satellite school—A "unique" educational experience for the EMR. *Journal for Special Educators of the Mentally Retarded*, 7(2): 82-85, 1971.

Project UNIQUE (United Now for Integrated Quality Urban-Suburban Education) is a new

educational effort designed to promote learning for all classes of children in Rochester, New York. Major components of the program include a demonstration classroom, community participation in education, work of teachers with mothers as aides in helping ghetto preschool children, teacher internships in inner city schools, financial aid, busing, and urban education courses for teachers. The objectives of the demonstration classroom are to display quality integrated education, to acquaint teachers and students with technological aids for teaching, to show the public a modern classroom, and to demonstrate innovative teaching techniques. Current federal financial support of Project UNIQUE may not be renewed, and the program will be able to continue only with public and private assistance. (No refs.) - *B. J. Grylack.*

Office of Special Education
Boces, Spencerport, New York

2227 Trainable mentally handicapped children learn to live in the community. *Deficience Mentale/Mental Retardation,* 21(1):26-27, 1971.

Ontario communities have been helped to become aware of TMR children and to evaluate and accept changes by the fact that schools for the retarded have become the responsibility of the Boards of Education. An applicant to a program is given careful consideration, and plans are made to meet the needs of the child through specific programs and to secure his rights. Applicants must have had a psychological assessment, must be toilet trained, and should be between the ages of 5 and 18 years. The main goals of the program include self-help, awareness of others, and the eventual ability to become a happy member of the community. Communication, arts and crafts, physical education, and living skills are stressed in the developmental curriculum. After age 18, the young retardate should be able to participate in a sheltered workshop program, where he will improve his social and physical skills and will undertake work assignments. (No refs.) - *B. J. Grylack.*

2228 HARPER, ROSEMARY; CLEARY, A.; & PACKHAM, D. An automated technique for the training of retarded children. *Programmed Learning,* 8(1):1-9, 1971.

Sixteen severely MR children were trained in matching to sample behavior with an automated training system, the touch tutor. A simple program containing 4 classes of stimuli (color, pictures, shapes, and words) and a more complex program in letter discrimination were used. Twelve of the children were trained to criterion on the simple program. Pictures, colors, shapes, and words could be sharply differentiated in that order of difficulty. On the complex program the MR children showed errors similar to those of normal children. The success of the system has implications for assessment as well as training. (11 refs.) - *E. L. Rowan.*

University of Newcastle upon Tyne
Newcastle upon Tyne, England

2229 DELACATO, CARL H. Neurological research in reading. In: Delacato, C., *The Treatment and Prevention of Reading Problems.* Springfield, Illinois, C. C. Thomas, 1971, p. 10-14.

This survey of experimentation leading to a neuropsychological rationale for treatment and prevention of reading retardation presents a theory of cortical dominance, but leaves diagnosis of reading difficulties still problematic, since cases with indications of psychological trauma, in which neurological evaluations are normal, are probably not of traumatic etiology, but rather of functional, organizational, or developmental neurological etiology. These cases, representing 70% of reading-retardation cases seen in most clinics, are the most difficult because there are no genuine techniques for evaluating them. (No refs.) - *B. Berman.*

2230 THOMAS, ELIZABETH C.; & YAMAMOTO, KAORU. School-related perceptions in handicapped children. *Journal of Psychology,* 77:101-117, 1971.

Examinations of attitudes of MR, disturbed, blind, and deaf children (CA range 10 to 16 yrs) toward 4 people and 4 subjects in the curriculum revealed a more heterogeneous reaction to people than to curricula and suggested that teacher preparation programs should differ for each group. The groups were not monolithic in their responses, and all responses reflected significant

handicap effects as well as interpersonal difficulties. The parent received the first position on all attitude ratings. Teachers were ranked second on movement and merit and last on security. Self was ranked third and classmates fourth. Self-esteem decreased with age. Science and language received first positions in the curriculum areas, and social studies received low ratings, especially among the MR, blind, and deaf. These data suggest that mental health in the classroom, current curriculum structure, and teacher preparation programs should be investigated. (23 refs.) - *J. K. Wyatt.*

Children's Medical Center
Tulsa, Oklahoma 74105

2231 **IHSAN, AL-ISSA.** Factors in the verbal and motor learning of imbeciles. *Journal of Psychology*, 77:151-156, 1971.

MRs from different age groups learned simple motor and verbal tasks and benefitted from this experience when learning related material. When MRs could not perform a simple motor task and then observed its completion by the experimenter, they tended to benefit from the observation. However, when groups of MRs observed the experimenter in the performance of a miniature task which was similar to the original task, a large number could not apply the experience to the solution of the original problem. When they performed the miniature task after the observation, performance on the original task improved. Age did not affect performance on the motor task but was an important factor in the abstract definition of simple words. Older Ss used more abstract definitions and were better able to generalize than younger Ss. (5 refs.) - *J. K. Wyatt.*

University of Calgary
Calgary, Alberta, Canada

2232 **CUMMINGS, ANN.** Operant conditioning incorporated in experiment using Sight, Sound and Symbol kit. *Journal on the Handicapped Child*, 4(2):8-11, 1971.

The Sight, Sound and Symbol kit which uses music to help develop basic reading skills and operant conditioning techniques were incorporated in an experimental program to teach 3 preschool children the letters of the alphabet. The project's primary purpose was to find out if the program had any appeal to children and their reaction to it rather than to reach any conclusions about the program's probable effectiveness in teaching reading. Each child was studied individually. Rewards consisted of cookies, candy, and toys as well as praise. A daily diary of procedures and reactions was kept on each child. Reaction to the rewards varied in the 3 children. One, for example, had to have explicit and controlled rewards and sometimes refused them; another performed better if praised after every note played on the piano. All were disappointed when the lessons ended. Recognition of alphabet letters on flash cards provided a measure of progress at the end of the project. The 6-year-old could recognize and associate the letters of the alphabet. The two 5-year-olds confused several letters but could match and associate them on the piano keyboard. All could distinguish the letters in their name. Excerpts from the daily diary are presented. (No refs.) - *J. C. Moody.*

Lincoln County Schools
West Virginia

2233 **HANRAHAN, HARRIET.** The name of the game–Completion. *Journal on the Handicapped Child*, 4(2):16-17, 1971.

A game form of operant conditioning was used in a reading class of 15 hyperactive, highly distractible EMR boys to increase interest in their work materials, and to encourage self-discipline and completion of assignments. Since the game's goal was to complete all class work, the word "completion" was introduced as a new vocabulary word and its meaning discussed before introducing the game itself. Each student's game board has 6 squares representing the days of the week and his name. The weekday sections are color coded to match the different colored rings on a target of concentric circles also representing the days of the week. The game boards are posted on the bulletin board next to the target. After finishing the assigned work the pupil places a tab of paper in the pocket of the game board for the appropriate day. On Monday colored yarn is placed from each game board on the target ring for Monday. Each day the assignment is completed, the yarn is moved towards Friday's center ring. Incomplete work disqualifies the student from moving his yarn or placing a

tab in that day's square. The game was successful: the boys developed better work habits and became very interested in their work. The game can be easily adapted in many ways for other classrooms. (No refs.) - *J. C. Moody.*

Cabell County Schools
West Virginia

2234 **PUNTER, AGNES.** Making the best use of television. *Teaching and Training,* 9(1):9-14, 1971.

The television series "Up in the Attic," planned for primary school children who are not making satisfactory progress in reading, was used 4 times a week for 3 months with 10 MRs (average CA 12 years, average MA 6 years) who were ready for reading to help them learn to read and communicate. By enlarging on and modifying the follow-up work for each 15-minute program suggested in the British Broadcasting Company's teachers' notes, there was enough material to cover whole days of activities. The activities described ranged from use of flash cards and plastic letters to related art and music projects, and also included a trip to the zoo. All the children were greatly stimulated linguistically and increased their vocabularies. One child can read all but 12 words in the words and pictures reading book, another can read most of the 8 stories, and the rest can almost read the first story in the book. All understand what they are reading about. (No refs.) - *J. C. Moody.*

John Farmer Training Centre
Christchurch, Hampshire, England

2235 **CARTWRIGHT, CAROL A.; CARTWRIGHT, G. PHILLIP; & ROBINE, GERALD G.** CAI course in the early identification of handicapped children. *Exceptional Children,* 453-459, February 1972.

A self-contained college level computer assisted instruction (CAI) course developed at the Pennsylvania State University is designed to give regular class teachers of seemingly typical children the knowledge and skills necessary to identify children who have problems that interfere with learning. Of 114 college students enrolled in an introductory course in special education, 87 were randomly assigned to conventional instruction (CI) and 27 to the CAI version of the course. The CAI group received all instruction by CAI; the CI group received instruction in the conventional lecture-discussion mode. On a criterion test of the major terminal behavioral objectives of the course, the CAI students received significantly ($p<.001$) higher scores than the CI students. Moreover, the CAI students completed the course in an average of 12 hours less than the 37.5 hours class time allotted by the university for a 3 credit course. These results are probably attributable to the intense individualization of instruction possible in CAI. An opinion survey of the CAI students indicated that 25 had favorable attitudes and 2 were neutral towards this method of instruction. The course is currently being widely disseminated by means of a mobile CAI facility to inservice teachers who cannot return to the college campus for refresher courses. The cost-effectiveness ratio has been favorable to the extent that funds have been secured to develop more CAI courses in special education. (7 refs.) - *J. C. Moody.*

Pennsylvania State University
University Park, Pennsylvania

2236 **GOLDSTEIN, HERBERT.** Education of the physically and mentally handicapped. *International Rehabilitation Review,* 22(1):18-19, 1971.

The 1970's promise to be the decade for both productive exploration and consolidation of new knowledge on MR. Research in special education which for many years focused on the differentiation of handicapped children from so-called normal children, shifted in the late 1950's and 1960's to studies of the learning and behavioral disabilities of MRs as children rather than as medical types. While the concept of learning disabilities is still fluid, the notion of behavior modification has emerged, which focuses attention on manifest behavior and ways to modify it rather than on internal or structural conditions. Much research in progress points to the new directions special education may take. Researchers in Yugoslavia have developed devices to enable the blind and visually handicapped to hear aloud printed material they have written or are reading. In the United States a study is determining the effect of training parents in educational

techniques on the educational achievements of congenitally deaf preschool children. German studies reveal a very high incidence of learning disorders and behavioral disturbances among children from socially disadvantaged areas. In India and Tunisia research is being conducted on the training and employment of MR youth. (No refs.) - *S. Glinsky.*

Rehabilitation International
New York, New York 10017

2237 **RYBACK, DAVID.** Cognitive behavior modification: Increasing achievement using filial therapy in the absence of supervision. *Canadian Journal of Behavioral Sciences,* 3(1):77-87, 1971.

Standard token reinforcement procedures were used by a mother to treat an MR son (CA 13 yrs) for dyslexia. The mother received supervision for a 7-month period during which there was significant improvement in the son's reading ability. Supervision was then withdrawn, and she continued to work with the boy for 2½ months. During this period of independent treatment, the boy made 24,234 single word reading responses. He averaged 120 words of reading for each penny value of reinforcement. (33 refs.) - *J. K. Wyatt.*

Rohrer, Hibler and Replogle
Montreal, Quebec, Canada

TREATMENT AND TRAINING ASPECTS — Psycho-social

2238 Editorial. *Journal of Mental Subnormality,* 20(2):55-56, 1970.

The principle of normalization suggests that MRs who have normal opportunities for living, experiencing, making choices, assuming responsibility, and working will be encouraged and stimulated to function at a higher competency level than if these opportunities are not available to them. These normal conditions need to be designed to compensate for the specific handicaps of individual MRs. The development of an integrated service for MRs will require national planning on a multidisciplinary basis. Redesigned institutional programs can provide stepping stones to normalization and assume a positive role in the development of this integrated service. (No refs.) - *J. K. Wyatt.*

2239 **KNOWLES, B. A.** Behavior modification and special education. *Slow Learning Child,* 17(3):170-177, 1970.

Behavior modification, which has affected most areas of human behavior with generally successful results, requires (1) careful specification of the behavior to be changed, (2) collecting data relevant to the behavior's occurrence, (3) manipulation of environmental events in a desired direction, and (4) additional changes, if desired effects are not achieved. Behavior modification begins with overt behavior and utilizes education to establish desirable patterns. Critics claim it is superficial, symptom-oriented, ignores inner causes of neurosis, and has only temporary effects; proponents deny this and claim permanent results. A number of workers have found behavioral principles very useful with the MR, and while it should not be regarded as a panacea, present indications suggest that it can be used to remedy problem behavior and to accelerate or improve academic performance in special education classes. (23 refs.) - *B. Berman.*

Department of Education
Western Australia

2240 **JENNINGS, A. N.; BALANDIN, D.; & MILLS, MILLIE.** Sex and the mentally retarded. *Rehabilitation in Australia,* 7(4):15-18, 1970.

Australia lags behind the more progressive countries in thinking and planning for the lives of adult MRs. MRs have as much right to sexual satisfaction as other people. Studies in the

United States and Denmark indicate that some MRs can marry and that many of the mildly MR can quietly assume normal patterns of living after marriage. In planning for the adult lives of MRs, it is important to consider their slower maturation process and the mismanagement, irritation, and confusion which may have arisen from mishandled sexual problems. Community living facilities for adult MRs are needed in New South Wales. In general the sex drive in MRs is lower than in normals, and the most severely handicapped may not even reach the first stage of sexual development. (3 refs.) - *J. K. Wyatt.*

Department of Health
New South Wales, Australia

2241 **KEYS SMITH, G.** Sex and the young sheltered workshop employee. *Rehabilitation in Australia,* 7(4):11-14, 1970.

The necessary functions for the physical side of sex life are present unimpaired in the vast majority of handicapped people, but the effects of some types of handicaps, especially paraplegia, can have a severe and devastating impact on the sex life. Handicapped persons often have bizarre ideas regarding the physical side of sex and may be very immature in their approach to sex. Marriage rates for sheltered workshop employees are very low. When marriage is considered, it is important to have accurate knowledge regarding disabilities, genetic complications, and possible physiological and adjustment difficulties. Sexual abnormalities may arise from feelings of bitterness and resentment or from feelings of inferiority and uselessness. The handicapped need to be helped to develop more mature personalities and assurance that they are respected. (No refs.) - *J. K. Wyatt.*

Royal Children's Hospital
Melbourne, Australia 3052

2242 **BHAGAT, M.; & *FRASER, W. I.** The meaning of concepts to the retarded offender. *American Journal of Mental Deficiency,* 75(3):260-267, 1970.

The technique of semantic differential, hitherto used primarily with normal intelligence populations, can be used to measure attitudes in MRs. The usefulness of the semantic differential with MRs was appraised, the attitudes of MRs exhibiting behavior disturbance were compared with those of average intelligence offenders, and it was ascertained whether this technique amplified interviews carried out by another investigator. The Ss were 81 institutionalized MRs who had severe behavior disturbance records and possessed a mean IQ of 65 and a mean CA of 29 years. These MRs were divided into 4 groups based on degree of rehabilitation, and were then compared with a group of 40 intellectually average young offenders. The Ss were given a test with 14 concepts relevant to delinquency, which were scaled according to the evaluative, potency and activity factors. A short-term test-retest on 25 Ss indicated the reliability of the instrument. The results showed that the evaluative and potency responses of the MRs and normals were significantly correlated, while activity responses showed poorer correlations. The findings suggest that the semantic differential is a useful instrument with qualified potential for measuring the attitudes of MRs of a wide range of intelligence. (10 refs.) - *S. Glinsky.*

*Lynebank Hospital
Dunfermline, Fife, Scotland

2243 **SHEPHERD, JOCELYN.** A message of good cheer from Laurence and his mother. *Intellectually Handicapped Child,* 9(4):18-20, 1970.

A mother of an autistic child relates the hardships, lack of official concern, and inadequate private help her family experienced in the care and development of their child. At each successive manifestation of autistic behavior little or no help was available from public, medical, or official sources. The child was helped primarily by the devotion and patience of the family and by volunteers, such as a play therapist, a music instructor, swimming instructors, a Sunday School teacher and, lastly, the Intellectually Handicapped Children's Society. The attitudes of educators, medical men, and public officials remain essentially apathetic and negative toward the autistic child. (No refs.) - *S. Glinsky.*

No address

2244 **AXELROD, SAUL.** Token reinforcement programs in special classes. *Exceptional Children,* 37(5):371-379, 1971.

When token reinforcement programs have been used in the classroom, the results have generally been positive. Programs have been used to increase academic performance of SMRs and multiply handicapped MR teenagers. A combined program in which token reinforcement was used with programmed materials resulted in increased studying and work accomplished as well as in reduction of disruptive behaviors. Such programs have also been successfully used to reduce hyperactive behavior, obscene conduct, and maladaptive behavior in MRs. A methodological problem in these studies has been the failure to clearly demonstrate that the contingent token reinforcement was responsible for the changes that occurred. Future studies should find methods to withdraw tokens without interrupting progress and should make greater use of reinforcers already present in the classroom. (19 refs.) - *J. K. Wyatt.*

University of Connecticut
Storrs, Connecticut

2245 **LARSON, THALIA.** Communication for the nonverbal child. *Academic Therapy,* 6(3):305-312, 1971.

A simultaneous sensory approach based on the language of signs used with the deaf is used along with traditional language stimulation methods to develop communication in nonverbal MR and/or multihandicapped children. This manual-oral method may prove successful with children with hearing loss, expressive motor problems, severely handicapping cerebral palsy, MR and physical conditions which prevent verbalization, and developmental delay. Sequencing in the development of sign language is similar to that in speech development. Sign language is not intended to replace but to stimulate the development of oral language. (No refs.) - *J. K. Wyatt.*

Special Education Schools
Orange County, California

2246 **MURRAY, M.** Rehabilitating the mentally subnormal. II. Social training of higher potential subnormals. *Nursing Mirror,* 132(3):44-45, 1971.

Although the behavior of MRs is accepted in the hospital, social training is needed to help them obtain greater acceptance in the community. MRs need to be aware of themselves as persons, and social training should emphasize the development of self confidence. Among the other areas which should be stressed in social training are care of personal appearance, deportment and control, experience in real life situations, development of independence, use of leisure time, and acceptance of criticism. (No refs.) - *J. K. Wyatt.*

Monyhull Hospital
Birmingham, England

2247 **SWARTZ, JON D.; DREW, CLIFFORD J.; & CLELAND, CHARLES C.** Facilitating mental retardates' social adaptation through corrective habilitation. *American Corrective Therapy Journal,* 25(1):16-18, 1971.

Habilitative training can improve MR social behaviors which traditionally stigmatize the MR. Two areas of deviant MR behavior which can be corrected are those of deficient posture and lack of facial expression. Poor posture may be improved through use of a meal-time table which can be gradually elevated and the employment of posture models viewed in films. Such modeling also may be used to improve the typical shuffling gait of MRs, which can further be helped by stepping exercises and marked footsteps on the floor. Facial training may be effected through taste, especially of citrus fruits, and odors, both of which cause marked facial expressions. These methods can engender other similar MR habilitative training techniques and lead to the development of new instructional materials. (10 refs.) - *S. Glinsky.*

University of Texas
Austin, Texas

2248 Mod-L Project—A token economy program in a remedial setting. *Deficience Mentale/Mental Retardation,* 21(3):19-20, 1971.

The Mod-L Project utilizes a token economy as a behavior modification approach in an attempt to provide a structural setting for an experimental school program for MR children with behavior problems. The project functions as a direct service to the retarded children in the class and

furnishes a framework for a variety of developmental research activities. The program is designed to handle from 8 to 10 children at one time. The primary criterion for admission is actual or near demission from regular school programs. Referrals are generally provided by the teacher or principal in the regular school. Participants in the program include the behavior therapist in charge of program operations, the teacher, and approximately 6 student trainees who serve on an alternating schedule as teacher aides and behavior raters. Within the classroom, emphasis is on the training and acceleration of appropriate behavior. In addition, the program develops a continuity in management techniques between the classroom and the home. (No refs.) - *B. J. Grylack.*

2249 **WEINER, LAWRENCE H.** New concepts in educating the emotionally disturbed retarded child. *Journal for Special Educators of the Mentally Retarded*, 7(2):106-110, 1971.

Recent aims in the education of children who are EMR and emotionally disturbed have been directed towards a daily program within the confines of the public school building instead of towards residential placement or other means of removing the child from the neighborhood school. Both reconstructed reality and reinforced reality are essential to the formulation of a program for the disturbed, retarded child. The teacher is concerned initially with reforming, clarifying, and redefining the interpretation of the environment by the child. Subsequently, the concept of reinforced reality is emphasized to ensure that only appropriate behavior is retained and that inappropriate behavioral responses are eliminated. A common sense approach which extracts some degree of interpersonal involvement by the teacher with the class, with emphasis on rapid change in the child's behavior and on the removal of problems of response patterns that interfere with learning, embodies both of these concepts. (No refs.) - *B. J. Grylack.*

Special Education Department
Barrington, Rhode Island

2250 **GARDNER, WILLIAM I.** *Behavior Modification in Mental Retardation: The Education and Rehabilitation of the Mentally Retarded Adolescent and Adult.* Chicago, Illinois, Aldine/Atherton, Inc., 1971, 379 p. $11.75.

The concepts and practices of behavior modification as applied to the treatment and education of the MR are examined. Emphasis is on the alleviation of those particular behavior problems which pose special difficulties for institutional staff. Taking into consideration the specific environmental and behavioral limitations of the MR in an institutional setting, a general analysis of learning processes is suggested. Individual sections are presented on respondent, operant, and observational learning, and examples of the functional approach to evaluation are discussed. A lengthy description of an existing research program for the design and testing of a sheltered workshop is also included as a model suitable for development by rehabilitation personnel generally. - *N. Mize.*

2251 **BUCHER, BRADLEY; & KING, LARRY W.** Centralization of punishment effects in the deviant behavior of a psychotic child. *Behavior Therapy*, 2(1):68-77, 1971.

Punishment with brief electric shock under various environmental conditions was used to suppress a form of destructive behavior in a nonverbal psychotic boy. A series of experimental situations which included differences in presession experiences, objects presented, persons present during the session, timing of the shock delivery, and experimental rooms were used to control touching of a forbidden object. When experimental conditions were changed, the punishment response was generally renewed. Suppression occurred with fewer punishments as the number of conditions increased. Punishment procedures should be consistently applied to produce suppression for nonspecific stimulus conditions. (10 refs.) - *J. K. Wyatt.*

University of Western Ontario
London 72, Canada

2252 **BERKOWITZ, SAMUEL; SHERRY, PATRICK J.; & DAVIS, BARBARA ALPHIN.** Teaching self-feeding skills to profound retardates using reinforcement and fading

procedures. *Behavior Therapy*, 2(1):62-67, 1971.

Behavior modification procedures were used to develop self-feeding skills in 14 institutionalized PMR boys (CA range 9 to 17 yrs; MA range 1.3 to 1.7 yrs) who had never spoon-fed themselves. The task was divided into 7 discrete steps and as each step was achieved by the S, the aide faded out that portion of her help until the S was making the entire self-feeding movement independently. All 14 developed self-feeding skills, and 10 were still feeding themselves 41 months later. (10 refs.) - *J. K. Wyatt.*

Crownsville State Hospital
Crownsville, Maryland 21302

2253 **BLAKE, SUE.** Operant conditioning paid off. *Journal on the Handicapped Child*, 4(2):12-15, 1971.

A program of operant conditioning successfully reduced the incidence of disruptive behavior in a special education class of EMRs. Class discussion of the behavior problems led to a solution of awarding points each day as rewards for not arguing or fighting with classmates, staying seated at one's desk, neater work sheets, better attendance, and better manners, e.g. not talking loudly without permission. A large chart listed each rule and the number of points for complying. Paper geometric figures with numbers represented the value of each reward. Individual envelopes for each child held the points collected every day. One hour every Friday was "spending time" during which the children used their accumulated points to rent such items as games, puzzles, autoharp, or record player and/or to buy drawing paper, balloons, candy, cupcakes, etc. Reward points could also buy special secret activities with the principal, guidance counselor, librarian, or custodian. The pupils were eager to acquire points and tell everyone they met in the hall about the points they had earned that day. The secret activities were considered particularly desirable rewards. After several months every child in the class had improved in behavior, academic work, and attendance. (No refs.) - *J. C. Moody.*

Kanawha County Schools
West Virginia

2254 **ALEXANDER, LINDA.** A study in operant conditioning with a multiple handicapped child. *Journal on the Handicapped Child*, 4(2):6-7, 1971.

A blind and hard-of-hearing girl enrolled in a TMR class had a very short attention span, was unable to sit still for any length of time, and was difficult to teach until operant conditioning was used. Operant conditioning aided Barbara in learning to use tactile discrimination in stringing beads and making bead patterns. Reinforcement by affection, which had previously been used with this child, had proven unsatisfactory after a time. Because Barbara confused the round M&M candy rewards with the beads, her reward was changed to ice cream. After a week of being rewarded for stringing first 1 round bead, then 2 beads, then 3 etc., she progressed to stringing 6 round beads before being rewarded. Although Barbara instantly realized how to string the square beads, she refused to string all 6 square beads before being rewarded and was therefore progressively worked up to 6 square beads using the same reinforcement pattern used for the round beads. This also took 1 week. She began learning a pattern of 1 round bead then 1 square bead, but a holiday lapse in instruction necessitated 2 additional weeks of work to gain what Barbara had learned before the holiday. Learning to string a given pattern when the square and round beads were mixed in the same box took almost a month. Similar work with cylindrical beads and then with a pattern of cylindrical and round beads was then mastered. Eventually Barbara could reproduce any pattern given her with few mistakes and little dependence upon receiving her reward. (No refs.) - *J. C. Moody.*

Kanawha County Schools
West Virginia

2255 **HAYWOOD, SCOTT.** The right reinforcement. *Journal on the Handicapped Child*, 4(2):23-27, 1971.

An operant conditioning regimen helped a 13-year-old hyperactive mongoloid boy (MA 5.4 years based on Peabody Picture Vocabulary Test and Goodenough Draw A Man Test, but a higher level of social functioning) with a very short attention span remain seated at his desk and concentrate on his schoolwork for increasingly longer periods of time. Observation determined

that 10-minute periods of concentrating on assigned work would be a reasonable initial goal. For 1 week his interruptions during a 10-minute segment were counted twice a day and plotted on a graph. This base-line measure of behavior, which showed an average of 6 interruptions in a 10-minute period, provided a precise indication of results as the program progressed. During the first week of conditioning the boy was given a small piece of cookie for every 2 minutes that he worked at his seat on an assigned task. Although he grasped the idea behind the reward during the first afternoon, he then earned it only once that day. By the end of a week, however, he had worked up to an average of 4 interruptions for a 10-minute session. During the next 2 weeks the number of interruptions was further reduced only slightly, but he did work for longer periods of time. The fourth week was highly successful. Although interruptions were reduced to 1.2 during a 10-minute session, he daydreamed or was otherwise distracted while remaining seated. By the end of that week, however, he was working for the entire 10 minutes. The transfer from physical reward to a verbal one, to which he adjusted very easily, was completed during the fourth week. The reward is now held off until midmorning (1½ hours after starting work), although his attention span is hardly that long. He will now work for at least 10 minutes on an assigned task without supervision, leaving the teacher free to work with the other students. The effectiveness of the program apparently lay in the type of reinforcement chosen and the goal set forth. (No refs.) - *J. C. Moody.*

Putnam County Schools
West Virginia

2256 **McDONALD, IAN.** Behaviour modification. *Rehabilitation in Australia,* 8(1):15-17, 1971.

Basic concepts and techniques of behavior modification, which is derived from research on operant conditioning, are reviewed. Behavior modification as a method that can help selected handicapped pupils is seen here, not as an end, but as a refreshing breeze in the rather conservative field of special education. (No refs.) - *J. C. Moody.*

Queensland Spastic Welfare League
Queensland, Australia

2257 **BAUMEISTER, ALFRED A.; & FOREHAND, REX.** Effects of extinction of an instrumental response on stereotyped body rocking in severe retardates. *Psychological Record,* 21(2):235-240, 1971.

An increase in nonadaptive behaviors in SMRs occurred as a result of nonreward. The effects of the extinction of well-learned instrumental response were examined in relation to stereotyped rocking behaviors of SMRs. The results demonstrated an increase in nonadaptive stereotyped movements in some SMRs as one of the behavioral concomitants of nonreward of an instrumental response. (9 refs.) - *S. Glinsky.*

University of Alabama
University, Alabama 35486

2258 **MILLER, HAROLD R.; PATTON, MARY E.; & HENTON, KAREL R.** Behavior modification in a profoundly retarded child: A case report. *Behavior Therapy,* 2(3):375-384, 1971.

Specific behavior modifications were achieved in a profoundly retarded child. The S was a 7-year-old PMR boy. Three programs developed a self-feeding operant, appropriate responses to a verbal command, and a greatly increased ability to stand without support. Side effects included an apparent increase in his responsiveness to his environment and independent mobility through crawling or using a walker. Observational followup indicates that his behavior gains have been maintained in his home. (11 refs.) - *S. Glinsky.*

Marshall State School and Hospital
Marshall, Missouri 65340

TREATMENT AND TRAINING ASPECTS – Occupational

2259 Rehabilitation in Australia. Westhaven Association, Dubbo, New South Wales. *International Rehabilitation Review*, 21(3):8-9, 1970.

Within the past 13 years, an association has been developed to assist MR children and adults through a complex of services including a special school and a vocational rehabilitation training program. A sheltered workshop has been established for the training program. Due to the basically rural location of the constituents, work opportunities are often realized through farming activities. Recreational needs are met by allowing the trainees 1 week every 3 months for supervised horseback riding, swimming, hiking, and similar sports. With the aid of volunteer work and government subsidy, the program has done much to increase community awareness of the rights and abilities of the disabled simultaneously with training MRs for a better life. (No refs.) - *B. J. Grylack.*

2260 **FENDELL, NORMAN.** Team work experience for the mentally retarded. *Rehabilitation Literature*, 31(11):330-332, 1970.

Work study programs for retardates in high schools represent a significant development in rehabilitation and special education during the last decade. Team work experience, a central concept in facilitating the transition of the student from school to employment, furnishes groups of MR students with supervised work in a variety of community settings, with a vocational instructor taking responsibility for the supervision and training of work crews at the job stations. The primary goal of team work experience will always be to help the MR to adjust to independent living. There appears to be general agreement among specialists that this goal is best achieved through a work-oriented program which gives the individual a feeling of productivity and values conforming to the cultural norms. (5 refs.) - *B. J. Grylack.*

Manchester Public School System
Manchester, Connecticut

2261 **ROSENTHAL, BERNARD S.** Separation anxiety as a factor in rehabilitation. *Journal of Rehabilitation*, 36(4):19-20, 1970.

In the past decade, a growing number of sheltered workshops has begun to assist handicapped individuals who are retarded emotionally or mentally in finding suitable employment. The problem is complicated by the fact that most employers are reluctant to hire MR individuals and experimental patients, and they place great importance on successful interviews. Many of the individuals served by sheltered workshops have tremendous feelings of inadequacy. As a result of effective workshop programs, clients begin to gain confidence. However, clients may develop somatic complaints and often question their abilities when they leave the workshop to perform in another setting, and counselors must therefore be cognizant of these resistances when they make job referrals. A recent nationwide study has indicated that job placement and counseling should constitute the final phase of workshop training. Client views of personal accomplishments should be sought as indicators of their perception of movement towards employment, and their attitudes towards the advantages of job placement in industry over employment in the sheltered setting should be utilized as motivation for job placement in industry. (No refs.) - *B. J. Grylack.*

Cincinnati Jewish Vocational Service
Cincinnati, Ohio

2262 **HANSON, ROBERT M.; BURGER, HARRIET; & PHILLIPS, ROBERT.** Handicapped youths learn child care. *Rehabilitation Record*, 11(6):24-26, 1970.

A course in child care provided by the Fairview Center in Westchester County, New York, for

adolescents with physical, mental, or emotional problems aims at possible future employment in child care for these young people. Students learn about the stages of early child development, are trained in nutrition, and are encouraged to develop patience, responsibility, attention to detail, and good grooming and cleanliness skills. The program – staffed by a school nurse-teacher and a teacher's aide – selects 3 babies (about 6 weeks old) from the community who are given total care for the entire school year. Each course, which is 8 weeks long, begins with general theory and concepts, and then progresses to practical skills and the tasks involved in daily baby care. The students have been strongly motivated, have developed positive attitudes toward infant care (boys, especially, have shown tenderness and affection), and several have been employed as teachers' aides in classes for young children. (No refs.) - *B. Berman.*

Board of Cooperative Educational Services
Westchester County, New York

2263 **CALLANDER, D. J.** Role of the opportunity workshop. *Intellectually Handicapped Child*, 9(4): 12-14, 1970.

The opportunity workshops of the Intellectually Handicapped Children's Society are an efficient and reasonably sized vehicle in terms of helping people, staff size, and money. The workshop, by definition, provides for the rehabilitation of intellectually, socially, and/or physically handicapped persons. In particular the 2 main functions are to provide free vocational training for those able to achieve independence and to furnish continuing employment and social opportunities for those unable to achieve independence. Training in the workshop must be closely related to the individual's needs and potential. Finally, the workshop's obligations extend to follow-up care after a trainee has left the workshop and also to provision of various social services, such as outings, sports events, dances, and opportunities for physical exercise. (No refs.) - *S. Glinsky.*

No address

2264 **O'HARRA, EDWIN A.** Using pay to change mentally retarded students' work behavior. *Teaching Exceptional Children*, 2(4): 163-169, 1970.

The In-Class Work-Study program for MR high school freshmen, sophomores, and juniors at the Del Campo High School, Fair Oaks, California, tested the effectiveness of operant techniques to teach work and social skills considered important to successful job performance and the advantages of using monetary payments as incentives for the acquisition of skills. Ten MR boys participated in a 20-week training program in a toy repair shop in a special classroom. The first shop session was unpaid, and the second, paid. All work in the simulated shop was done in a systematic manner, with the amount of money being paid to the students dependent upon 6 social and 6 work factors. The teacher and teacher's assistant rated each student's daily performance on a scale of 0 to 5 points, ranging from task avoidance to superior performance. The correlation between scores for most social and work factors was positive and high and varied from .83 to .97. The pay factor appeared to be critical in creating a successful simulation of a real work situation and made a significant difference in the quantity and quality of work production during the paid period, in which the overall production rate increased by 40%. (2 refs.) - *B. J. Grylack.*

Del Campo High School
Fair Oaks, California

2265 **MALIN, A.** Recent taxonomical trends in sheltered employment for the mentally retarded. *Journal of Rehabilitation in Asia*, 11(4):18-20, 1970.

It is crucial to distinguish between the terms "sheltered workshop" and "sheltered employment" in regard to MR individuals and to recognize the need for the latter, broader term, for the importance of sheltered employment is related directly to the significance which society attaches to the concept of labor. The 4 proposed major work or adult training program areas, delineated according to the nature and degree of mental impairment, include selective placement for individuals capable of functioning independently in semi-skilled and unskilled positions or even in skilled jobs, vocational rehabilitation and sheltered workshops, activity centers, and independent living centers. In this outline, the concept of vocational rehabilitation is limited to the sheltered workshop, however. A taxonomy must be found for outdoor non-shop occupations, which are becoming increasingly significant and

which can match suitably the controlled elements of the sheltered workshop. "Sheltered work farm" is the best term at the present time. (7 refs.) - *B. J. Grylack.*

No address

2266 **DINNISS, PETER E.** Taranaki Opportunity Workshop developments. *Intellectually Handicapped Child*, 10(1):13-17, 1970-1971.

Since the establishment of the Taranaki (New Zealand) Opportunity Workshop in a pleasant semirural environment with ample recreation area and space for future developments, approximately 68 handicapped trainees have participated in activities ranging from self-care training to complex construction and assembly tasks. At the present time, 41 individuals are enrolled. Most of the work is contract-based, but the amount of contract work undertaken is always carefully controlled to permit flexible programming and to prevent undue pressures arising from contract deadlines. Only about 60% of the 35-hour weekly attendance is devoted to paid work training and production. Almost 40% consists of social training. It has been the combination of these factors which has contributed to the success of the total program not only for those who have been rehabilitated and are now in outside employment but also for those who require continuous sheltered employment. Greater attention to the needs of trainees who are distractible and lack attention control is planned for 1971. (No refs.) - *B. J. Grylack.*

Department of Education
New Plymouth, New Zealand

2267 **MURRAY, M.** Rehabilitating the mentally subnormal. I. Occupational and social training. *Nursing Mirror*, 132(2):32-33, 1971.

Occupation for the MR is necessary for the development of self respect, learning, sublimation of frustrations, identification with the community group, and the satisfaction of physiological and psychological needs. Work provides an opportunity for the admiration and affection necessary for self respect. A well thought out program of occupation can increase social awareness, make up for inadequacies, and open the door into the community. (No refs.) - *J. K. Wyatt.*

Monyhull Hospital
Birmingham, England

2268 **GARRETT, JAMES F.** Economic benefits of programmes for the retarded. *Programs for the Handicapped*, 71(2):1-8, 1971.

Data from 3 rehabilitation projects indicate that economic benefit ratios for MRs are at least as high as the average for all rehabilitants and are probably higher. A study of earnings-cost ratios for selected client groups revealed a ratio of 4.65 to 1 for MRs, well above the average for all groups of clients (3.58 to 1). Only 9% of the MRs employed in the Federal employment program have been separated because they were unable to perform and/or adjust socially. Three groups of Federally funded demonstration projects represent an actual total of 80 projects. These projects have emphasized a direct educational, training, vocational, and placement thrust. Research has been practical and had a vocational orientation. Research and demonstration costs have been modest. The number of MRs who have been rehabilitated began to increase rapidly (106 in 1945; 26,762 in 1969) soon after the Federal government became involved in research. (No refs.) - *J. K. Wyatt.*

Social and Rehabilitation Service
Dept. of Health, Education, and Welfare
Washington, D. C.

2269 **JACKSON, JAMES L.** Extended rehabilitation services for the mentally and physically handicapped. *Rehabilitation Literature*, 32(2):43-44, 1971.

A pilot program created by the Texas Rehabilitation Commission will provide sheltered workshop employment opportunities, extended community living services, and other benefits to SMR individuals who are ineligible for vocational rehabilitation sponsorship. Purchased services include personal and social adjustment training, work adjustment training, on-the-job training,

room, board, and 24-hour supervision of retardates. Because these SMR persons are separated from their homes, considerable time is often required to orient them and their families to the pilot project. Funding for the services is requested annually from the state legislature. An administrator-coordinator will be responsible for the day-to-day program and will be assisted by 14 people placed in the major field offices across the state. (No refs.) - *B. J. Grylack.*

Texas Rehabilitation Commission
Austin, Texas

2270 **HENRI, JACQUES.** La conception des Centres d'Aide par le Travail (The concept behind Centers of Assistance through Work). *Nos Enfants Inadaptes,* 36:3-6, 1970.

In recent studies undertaken by the UNAPEI, the desirability of establishing a series of workshops for handicapped persons was underscored. A correct understanding of the concept behind workshops for retarded adults has to recognize, first of all, that the 'shops,' although not educational in the strict sense, contribute primarily to the overall development of the MR men and women employed there. Work has to be found which can be suitably adapted to the skills level of the retarded. However, at the same time, the products created should have commercial value, and should be able to compete effectively on the market without major financial support to the workshops from outside institutions. The shops should also take into account the different interests of the workers, ranging from wages and recreation facilities to supplementary education. Aside from the administrative structure of the particular shops, based on a director with supervising personnel, additional service-institutions (commercial, administrative, financial) should be available for larger groups of workshops. (No refs.) - *P. Custers.*

No address

2271 **JULIEN, CLAUDE.** Les handicapes mentaux (The mentally retarded). *Readaptation,* 177:47-51, 1971.

One of the problems facing rehabilitation centers in France is the adaptation of the training in these centers to the needs of the labor market. Often handicapped young persons who are discharged by the Department of Social Security are unable to find a job and can be employed only in special workshops. An example is given of a Center in Paris which has been able to successfully reconcile the exigencies of the individual reeducation process with the possibilities of employment afterwards, through 'contracts of rehabilitation'. The re-education center functions as an intermediary with the employers and after the handicapped are employed defends their interests according to terms of the contract. Both men and women are enrolled in the program. (No refs.) - *P. Custers.*

Office of Social Protection
Paris, France

2272 **CORVIN, GERDA.** "Tell him." Part II. *Journal for Special Educators of the Mentally Retarded,* 7(2):115-118, 1971.

In a good counseling relationship, the client should be able to discuss almost anything that goes on within him without shame or embarrassment with the knowledge that the counselor is genuinely interested and sympathetic. This kind of experience has proved unique for retardates who had been largely ignored. In the sheltered workshop for the retarded, trainees are assigned to counselors as a means of dealing with personal family problems which affect their work habits and other social adjustments. The counselor aims to reduce excessive anxiety, insecurity, depression, and rebelliousness, and to help the client become more self-reliant and self-aware. The counselor must keep pace with his client, whether it is a slow or fast one. Cooperation on the part of parents is often required to accomplish counseling goals. (No refs.) - *B. J. Grylack.*

A.H.R.C. Workshop
380 Second Avenue
New York, New York 10010

2273 **NITZBERG, JEROME.** Functions of sheltered workshop supervisors (shop teachers). *Journal for Special Educators of the Mentally Retarded,* 7(2):86-89, 1971.

Sheltered workshop supervisors function as teachers of work skills that are appropriate for retardates. In addition, it is their responsibility to help

evaluate trainees, to establish some limited contact with parents, to assure the availability of work, to act as members of the habilitation team, to conduct idle time programs, to participate in workshop inservice training programs, to help to explain the workshop to visitors, and to strive to develop in trainees proper work habits, attitudes, and relationships with peers and authority figures which are necessary for the performance of mature and competent workers. Training center supervisors perform all of these tasks, but they have greater responsibility in such areas as evaluation, habilitation, team work, and individual client relationships. Senior branch workshop supervisors fulfill the functions of the workshop supervisors in addition to managing branch workshops, a task which requires performance of administrative, maintenance, supply, receiving, shipping, and clerical duties. (No refs.) - *B. J. Grylack.*

A.H.R.C. Workshops
New York, New York

2274 **BROWN, ROY I.** Development handicap and the Vocational & Rehabilitation Research Institute (symposium). *Deficience Mentale/Mental Retardation*, 21(1):21-24, 1971.

The Vocational & Rehabilitation Research Institute in Calgary, Canada, assesses, monitors, and evaluates all of its programs as the services are delivered to developmentally handicapped persons. More emphasis is placed on social knowledge, emotional stability, and adaptability than on cognitive performance in predicting an individual's success. New trainees are placed either in the training and assessment laboratory, which deals with simulated industrial production and assembly of simple goods, or in an industrial workshop. Achievement of high levels of motivation is influenced by the realistic and concrete goals which are established for the individual. The Institute's residence for trainees provides instruction in home living and social behavior within the community. When an individual approximates the performance of an average worker in the community, he is referred to the placement committee. Parental counseling is an aspect of training which has not yet been fully developed. (No refs.) - *B. J. Grylack.*

University of Calgary
Calgary, Alberta, Canada

2275 **FLAME OF HOPE, INC.** *Flame of Hope: New Approaches to Competitive Employment of the Mentally Retarded through Proprietary Products. Final Report.* Hyannis Port, Massachusetts, Flame of Hope, Inc., 1971, 110 p. (Mimeo)

The Flame of Hope Project, a network of 13 sheltered workshops throughout the United States, has successfully employed over 750 MR persons in the commercial production of candles and perfume. Established in 1968, the project has been supported by the U.S. Social and Rehabilitation Service and by matching funds from the Joseph P. Kennedy, Jr., Foundation. The wholesale value of the products produced during this period has been well over $1 million. Quality control and management responsibilities are handled by workshop staff members. By deducting a marketing fee from the products sold, Flame of Hope, Inc., has become economically self-sufficient, and it is expected that the project will rely solely on this income source for financial support in the future. The success of this sheltered workshop program can be measured both by the achieved integration of rehabilitation and training for the MR clients with the production of saleable merchandise, and by the consistently high levels of production, quantitatively and qualitatively, which indicate that this project might usefully serve as a model for future efforts in the field of occupational rehabilitation for the MR. - *N. Mize.*

2276 **RUSALEM, HERBERT; BAXT, ROLAND; & MILLER, ALFRED P.** *Vocational Rehabilitation of the Mentally Retarded Adult: Report of a Demonstration Project Featuring an Inter-Generational Relationship.* New York, Federal Employment and Guidance Service, 1971, 55p.

A specialized vocational rehabilitation program was able to assist MR adults who had repeatedly failed in the labor market to remain as autonomous individuals and continue to function in the community. An important aspect of this demonstration project was the buddy system, where another disabled person assumed specific responsibilities relative to the MR client. (55 refs.) - *E. L. Rowan.*

215 Park Avenue South
New York, New York 10003

2277 **JOHNSON, GIL; & TUTTLE, DEAN.** Education and habilitation of multiply handicapped blind youth. *New Outlook for the Blind,* 65(2):56-61, 1971.

The California School for the Blind and the State Department of Vocational Rehabilitation provided services for adolescent multiply handicapped blind students. Handicaps in addition to blindness included MR and cerebral palsy. Programs were provided for a high school group, a transitional work experience group, and a work evaluation group. Individual achievements varied. Although all students could not attain independence and self reliance, accomplishments justified the expense of the program. Group services and capabilities for each group are presented in detail. (No refs.) - *J. K. Wyatt.*

No address

2278 **WEINBURG, M. M.** The adolescent away from home. *Teaching and Training,* 9(1):23-24, 1971.

Pengwern Hall, which caters chiefly to boys and girls from 15 to 21 years old within an IQ range of 46 to 100, has for the past 3 years conducted experimental training courses in various job skills. The 3-month courses entail a work program based on a 5 day/40 hour week. The trainees work alongside the staff which gives opportunity to teach a sense of responsibility. The boys work on the 22 acres of grounds or in the greenhouses in order to learn skills for potential employment as simple manual laborers, building laborers, gardeners, assistants for horticultural work, etc. Eventual canteen or laundry jobs are envisaged for the girls. Regulations are minimal, the major one concerning regular bedtime and arising hours so that trainees will arrive at work on time. Trainees are encouraged to spend their leisure time in the nearby town swimming, dancing, or at the cinema. They are also encouraged to entertain other young people at Pengwern Hall as they would at home. In addition pottery classes, squash, and other activities are available. The success of the courses has varied greatly. Some youngsters have had success in finding work, some have not, and some have had a great variety of jobs since leaving. It has been found that the younger the trainees are when they enroll, the greater is their chance of achieving success. Pengwern Hall also offers holidays and short term care for the mentally handicapped. No fewer than 4,000 children have been received in the last 3 years. (No refs.) - *J. C. Moody.*

Pengwern Hall
Nr. Rhyl, North Wales

2279 **POSNER, BERNARD.** They did not see me. *Rehabilitation in Australia,* 8(1):28-29, 1971.

Accompanied by a young moustached rehabilitation counselor, the author posed as a MR worker seeking a job. After 2 days spent calling on a dozen employers of all kinds in a western U. S. city, it was found that (1) there was no out-and-out rejection because of being retarded; (2) there was no doubt that as an MR he was able to work at something; and (3) there was no doubt about the kinds of lesser skilled work he was able to do. The experience was degrading and dehumanizing, however, in that most potential employers talked about the MR job seeker in his presence as if he were not there. Even basic questions were addressed to the rehabilitation counselor. (No refs.) - *J. C. Moody.*

No address

2280 Hospital work stations for MR's. *Rehabilitation Record,* 12(2):20-21, 1971.

A work-study program for EMRs in a hospital prepares them for future occupations outside of sheltered workshops. Thirteen high school sophomore EMRs, with IQs between 50 and 75, work one 5½ hour day per week at the Downstate Medical Center in Brooklyn, New York. The EMRs work in X-ray, records, outpatient clinic, pharmacy, and receiving, and go to and from work independently by public transportation. As the program has been highly successful, paid summer jobs as well as eventual full-time employment are contemplated for the EMRs. The work-study program has educated not only the EMRs but the public as well. (No refs.) - *S. Glinsky.*

2281 **DOLNICK, MICHAEL M.** Sheltered workshop programs in the Netherlands. *Rehabilitation Record,* 12(2):35-38, 1971.

The social workshop program in Holland is extremely successful due to cooperation between the central government, labor unions, and industry. The sheltered workshop program in Holland is based on the 1969 Social Employment Act, which provides for 180 social workshops employing 44,000 persons, or 1 out of every 320 in the country. The United States' involvement is only one-seventh of this. The underlying concept is that the government is responsible for creating sufficient employment for every citizen. Citizens unable to enter regular employment work either in social workshops, outdoors projects, or white collar projects. The government annually budgets 1¼% of its national budget for social workshops, which are large and resemble industry as closely as possible. Employment eligibility requirements include the ability to do productive work at one-third the rate of a normal worker, age less than 65, and Dutch citizenship. The wage system gives the worker 95% of the minimum wage and an opportunity for up to a 20% bonus. A second aim is vocational rehabilitation: every year about 6% of the workshop population leaves for regular industry. The social workshop concept is rooted in the Dutch belief in the therapeutic value of work. (No refs.) - *S. Glinsky.*

No address

2282 **SLATER, NATHAN M.; & FOX, JUDITH.** Readin' plus 'rithmetic equal a new image. *Rehabilitation Record,* 12(2): 26-29, 1971.

Remedial basic education in the Federation Training Services Program has provided minimal basic skills relevant to specific vocational goals. Reading and arithmetic are recognized as critical vocational skills, and training is provided in these areas to clients of the Federation of the Handicapped in New York City. Remediation has been given in preparation for the vocational areas of food service, electronics wiring and assembly, mechanical inspection, porter-building maintenance, and mailroom services. The training is functional, related to actual work skills, and continues for 12 weeks. The training classes in reading are open-ended and composed of small groups, while mathematics classes are on a one-to-one basis. Specifically oriented technical training is supplied after achievement of basic reading and mathematical objectives. Several successful placements have been made and additional benefits realized, such as the sustaining of trainee interest during training, the acquisition of life skills, and the enhancement of self-concept through successful work in the remedial area. (5 refs.) - *S. Glinsky.*

No address

2283 **SONG, R. H.; & SONG, A. Y.** Development of a vocational adjustment rating scale for the retarded. *Journal of Counseling Psychology,* 18(2):173-176, 1971.

A highly reliable vocational adjustment rating scale for MRs has been developed. One hundred thirteen MRs were utilized as Ss in developing the scale, which is composed of 52 brief items and showed high reliability and concurrent validity. The results support the hypothesis that MRs classified in supervisors' ratings as better workers can be significantly different from those classifed as poor workers. Further research to standardize the scale is recommended. (6 refs.) - *S. Glinsky.*

Wisconsin State University
Whitewater, Wisconsin 53190

TREATMENT AND TRAINING ASPECTS – Therapy

2284 **FELL, J. H.** Drama and the subnormal patient. *Nursing Mirror and Midwives Journal,* 131(14):36-37, 1970.

The value of drama as a means of educating, rehabilitating and training subnormal patients is being appreciated more and more. While musical productions have the greatest value, "straight" plays and mime for the very retarded result in excellent gains for the participants. Best results have been obtained where the plays have been specially written to best display the patients'

capabilities, and where costumes, scenery, stage properties and make-up are in generous supply. (No refs.) - *G. S. Bell.*

Calderstones Hospital
Whalley, nr Blackburn
Lancashire, England

2285 MELIN, BENGT. Observations on the use of Beclamide (Nydrane) in a child psychiatric clinic. *Journal of Mental Subnormality*, 16(31):119-120, 1970.

Beclamide (N-benzyl-beta-chloropropionamide) was effectively used in many children for the treatment of aggressive behavior, psychomotor anxiety, suspected cerebral damage, and/or tics. Although the intelligence of the 40 patients (CA range 7 to 18 yrs) was within the normal range, many of their symptoms were similar to those observed in MRs. Of 33 children with the main symptom of aggressiveness, 17 improved with Beclamide treatment alone, and 2 with Beclamide in combination with other drugs. Ten of 28 children with psychomotor anxiety and restlessness improved with Beclamide alone, and 5 improved under combined treatment conditions. In 9 children with suspected cerebral damage, improvement after treatment with Beclamide was noted in 5, and 1 of 3 children with tics evinced dramatic improvement when the drug was used. Improved conditions were maintained over a long period of time, and no side effects were observed. (2 refs.) - *J. K. Wyatt.*

Kalmar Hospital
Kalmar, Sweden

2286 SHENNAN, VICTORIA. Dr. Kassorla speaks . . . *Parent's Voice*, 2(4):10-11, 1970.

Operant conditioning principles were used to produce acceptable behavior and increased communication skills in a 12-year-old autistic girl. In a 2-month-long intensive treatment program which involved over 60 people, constant therapy sessions were provided for the child. Acceptable behavior was rewarded with praise, sweets, and/or affection, while the therapist's response to psychotic behavior was rejection or disapproval. This therapy is not considered to be a cure for autism but does bring about behavioral changes which make life easier for those who live with autistic children. (No refs.) - *J. K. Wyatt.*

No address

2287 GREEN, ELIZABETH A.; & TAYLOR, RODNEY H. The eccentric mattress. *Parent's Voice*, 29(4): 22-23, 1970.

A therapeutic mattress composed of foam shapes covered with neutral-colored, waterproof, spongeable material of a stimulating texture is designed for play use and to increase investigation of the world by children with any handicap, but especially for spastic children, mongols, and children with perceptual disturbances. The 5 basic shapes can be used in physiotherapy, social training, and free play. (No refs.) - *J. K. Wyatt.*

Cell Barnes Hospital
St. Albans, England

2288 BICKNELL, JOAN. Tall in the saddle. *Mental Health*, Autumn:29-31, 1970.

The provision of horseback riding facilities for the handicapped allows these children to do something that normal children wish to emulate and creates a feeling of independence, mobility, and the realization that the animal is responding to their directions. The first effect upon a mentally or physically handicapped child when he begins to ride is psychological. Both hospitalized and home patients participate in the several riding groups that have been organized for handicapped children. Volunteer help is often essential to the successful functioning of these groups. (No refs.) - *B. J. Grylack.*

No address

2289 BAKER, BRUCE L.; & WARD, MICHAEL H. Reinforcement therapy for behavior problems in severely retarded children. *American Journal of Orthopsychiatry*, 41(1):124-135, 1971.

Six SMR children and 6 controls with IQ<25 and CA of 6 to 9 years were used to study the

usefulness of a behavior modification program utilizing reinforcement techniques in which the primary focus was elimination of behavior problems. The results were assessed by intelligence and developmental testing, independent behavioral observations, and records kept by the project staff. Treatment was carried out by selected ward personnel following a brief training period. The target children improved in stereotyped behavior and specific problems, such as temper tantrums or running away. These children also demonstrated improvement in responsiveness to instructions, play skills, interpersonal relationships, self-help and academic skills, and overall developmental level. It is not meaningful, however, to separate the milieu from reinforcement therapy, since the availability of reinforcers is essential to a total reinforcement therapy program. Reinforcement techniques are taught readily to nonprofessionals and thus encourage the expansion of available treatment. (23 refs.) - *B. J. Grylack.*

Harvard University
Cambridge, Massachusetts 02138

2290 **COLLINS, VIRGINIA.** Coordinated care of the child with spina bifida. *Lancet,* 2(7683):1140, 1970. (Letter)

Support is given to the practice of treating spina bifida patients in joint meetings of various staff personnel. Such treatment would save time, expense, and effort, require fewer visits, and would be much more convenient for the handicapped patient and his family. (1 ref.) - *E. Kravitz.*

Queen Mary's Hospital for Children
Carshalton, Surrey, England

2291 **MOFFATT, W. R.; SIDDIQUI, A. R.; & MacKAY, D. N.** The use of sulthiame with disturbed mentally subnormal patients. *British Journal of Psychiatry,* 117(541):673-678, 1970.

Forty-two SMR markedly abnormal psychiatric inpatients with severe behavioral disturbances were used as Ss in a study of the significance of sulthiame as a tranquilizer. A double-blind cross-over trial was designed, and results were collected on the basis of rating scales and observations by nursing staff. Sulthiame, which had been studied previously as an anticonvulsant, was found to be significantly effective in decreasing the incidence of behavioral disturbances, although it did not act selectively on different forms of deviant behavior. Sulthiame showed success with patients who had not responded favorably in an earlier trial with pericyazine and chlorpromazine. Side effects from sulthiame were absent in this study. (9 refs.) - *B. J. Grylack.*

Muckamore Abbey Hospital
Co. Antrim, Northern Ireland

2292 **POPE, MICHAEL.** Art, play and slow learners. *Special Education,* 59(4):19-23, 1970.

The art teacher should develop a relaxed atmosphere and should use himself as a responsive instrument to create continuous order and purpose. The child should enjoy art and should develop imagination, observation, and appreciation. Projects should be challenging but should not be too difficult or time consuming. The teachers should motivate but not impose, look for creative insight, and move in the direction of the unknown. They must be aware of when to help and should be able to recognize creativity. The teacher should recognize individual temperament and ability and assign appropriate projects. (No refs.) - *J. K. Wyatt.*

Waltham Forest ESN School
East London, England

2293 **GORDON, J. G.** Speech therapy. *Intellectually Handicapped Child,* 9(4):8-10, 1970.

Understanding speech development in normal children enables parents of MR children with speech deficiencies to assist the child's advancement knowledgeably. Speech may be delayed in a child by a multiplicity of factors, including deafness, damage to cortical motor areas or pathways from the brain to the muscles, and disturbance in the sensory pathways or in conceptual functioning. Speech development in the child proceeds from initial listening, through understanding to talking. The child's general level of

development determines how quickly he advances, and an MR child or delayed child can be assisted by one's speaking in simple phrases, pausing between ideas, repetition, associating words with things and actions where possible, and in general structuring the world of words and ideas to accommodate his ability level. (No refs.) - *S. Glinsky.*

Thorndon School
Wellington, New Zealand

2294 **CARTER, JOHN L.; & MILLER, PHILLIS K.** Creative art for minimally brain-injured children. *Academic Therapy*, 6(3):245-252, 1971.

Eight minimally brain-injured children (age range 7.2 to 9.9 yrs; IQ range 68-108) who participated in a creative art program designed to provide learning incentive and the development of perceptual awareness and manipulative skills made significant gains in perceptual motor ratings. The class met daily for 2 hours for 5 weeks. Activities were designed to increase self-awareness, identification with experiences, eye-hand coordination, and interest span, and to improve self image. Pre- and post-test scores on the Frostig Developmental Test of Visual Perception evidenced a mean gain of 15 points and a mean perceptual age gain of 7.32 months, both of which were significant ($p<.05$). All gains for all children for each subtest were positive and significant ($p<.05$). (3 refs.) - *J. K. Wyatt.*

University of Houston
Houston, Texas

2295 **OBALDIA, MARIO DE; & BEST, GARY A.** Music therapy in the treatment of brain-damaged children. *Academic Therapy*, 6(3):263-269, 1971.

A music intervention program was used to educate and treat 10 brain-damaged children, 2 of whom were diagnosed as untestable and autistic. The program included group singing activities and involvement with specific musical instruments. All Ss showed improvement. One autistic boy (CA 10 yrs) who had suffered from violent rages exhibited almost immediate changes in behavior. Sociability and self assurance increased, self castigation decreased, and he became more docile and obedient. The second autistic boy (CA 10 yrs) was blind and his behavior was rebellious and violent. After involvement in the program, rebelliousness decreased, stubbornness diminished, and self-assurance increased. Music therapy appeared to provide a decisive intervention technique for the training and treatment of several categories of brain damage. (1 ref.) - *J. K. Wyatt.*

Penny Lane
Sepulveda, California

2296 **MENOLASCINO, FRANK J.; & OSBORNE, ROBERT G.** Psychiatric television consultation for the mentally retarded. *American Journal of Psychiatry*, 127(4):515-520, 1970.

A long-distance communication system was designed to provide psychiatric consultation and supervision to a group of 60 MR adults. Using a closed circuit television, nonresident specialists on the staff of the Nebraska Psychiatric Institute in Omaha gave psychiatric counseling to the patients and guidance to the resident staff at the Norfolk State Mental Hospital 112 miles away. Monthly 1-day visits by the specialists to the mental hospital were established to promote personal contact with the patients and resident staff. At the end of 6 months of television consultation, newly structured approaches were being employed at the mental hospital, and the number of volunteers had increased. The sequential program that this television-aided consultation service has produced for the MR patients encompasses individual assessment of the patients by the staff, expanded dimensions in the personal and social range, vocational assessment and placement, and gradual integration of the patients into the community at some productive social and vocational level or into a program at the mental hospital which fulfills the need for a sheltered living setting. (10 refs.) - *B. J. Grylack.*

University of Nebraska Medical Center
Omaha, Nebraska

2297 **LAHR, IRENE W.** Adaptation of walker for child with ataxia. *Physical Therapy*, 50(10):1490-1491, 1970.

A simple modification of an adjustable pick-up walker made independent ambulation possible for

a retarded child with ataxia. The small walker utilizes ball bearing casters on the front posts and is much more manageable for a very small child than the conventional 4-wheeled walkers. The walker has permitted ambulation of very small children with myelomeningocele and cerebral palsy who could not cope with crutches. (No refs.) - *B. J. Grylack.*

2814 Heritage Avenue N.W.
Canton, Ohio 47718

2298 FOX, SHEILA. When health is a handicap. *Mental Health,* Spring:29-31, 1971.

Since there has been no documented report of a cure for brain-damaged children, the Doman method offers unprecedented hope to parents who want this rigorous treatment—the impression of a pattern on the child's brain—administered to their children. However, the constant programming involved in the Doman method precludes any normal life for other members of the family, and it is difficult to say how long it would take to achieve the goal of a completely well child. While many people adjust amazingly well to having a handicapped child, others do not. If the situation is tragic for parents and child, it is no less traumatic when a normal brother or sister has a handicapped sibling. Unless parents are urged to consider the family as a whole when they decide upon a course of treatment for their brain-damaged child, the consequences can be severe. The future of the handicapped depends upon the concern and generosity of the present generation. Those with handicapped siblings will make significant contributions only if they have not experienced rejection in a family too involved with a handicapped child. (No refs.) - *B. J. Grylack.*

Spastics Society
Somerset, England

2299 LIBO, SHIRLEY S.; PALMER, CAROLYN; & ARCHIBALD, DOROTHY. Family group therapy for children with self-induced seizures. *American Journal of Orthopsychiatry,* 41(3):506-509, 1971.

In view of findings that epileptic attacks may be precipitated by sensory stimulation and that self-induction of seizures has been known to occur, the motivation of 2 photosensitive epileptic children for inducing seizures was explored. Both Ss were neurologically unremarkable, had low normal intelligence, had photosensitive EEGs, and had failed to respond to various anticonvulsant medications or to retain dark glasses. The Ss and their parents became part of a joint family group with a psychologist and social worker. As a result of initial success with this method, other epileptic children were included. When all significant family members were involved, supportive psychotherapy and mutual support of the group seemed to help the family to deal better with the child's symptoms. Behavioral control of the self-induced seizure is possible once the reinforcing consequences of provoked seizures are understood and families are able to alter their responses. (13 refs.) - *B. J. Grylack.*

Child Guidance Center
Albuquerque, New Mexico 87108

2300 CLINE, DAVID W. Video tape documentation of behavioral change in children. *American Journal of Orthopsychiatry,* 42(1):40-47, 1971.

Video tape was used to document behavior before therapy and at 4-month intervals during 1 year of psychotherapy with a pair of identical 7-year-old twin boys with childhood schizophrenia. A normal male peer was used for comparison. Each video-taped record consisted of a standardized 30-minute interview with the whole family plus another 30 minutes devoted to the twins alone together, each twin alone with the normal peer, and the twins together in play therapy with the child psychiatrist consultant. Two judges rated the tapes independently using a standardized behavioral coding system; overall agreement between the judges was 67.8%. Results show that behavioral change can be reliably documented both qualitatively and quantitatively using this technique. (16 refs.) - *J. C. Moody.*

Mayo Box 393
Minneapolis, Minnesota 55455

2301 MAXWELL, JANE E. Home care for the retarded child. *Nursing Outlook,* 19(2):112-114, 1971.

The public health nurse can be the prime catalyst in providing home care for MRs. The nurse's responsibilities in work with MRs encompass prevention, casefinding, and home management. Possible candidates for home care are selected on the basis of the parents' willingness to work with the child at home and the MR's potential for home training. The nurse's frequency of initial contact with the MR and the encouragement of the parents are essential to success. MR home training involves coordination with the mother in establishing the priority of the MR's needs, establishment of attainable goals, inventory of family and community resources for helping the child, initiation of a realistic program of scheduled activities, and regular evaluation of progress. One such program in Pomona, California, has resulted in a dramatic increase in MRs receiving home as opposed to institutional care. The public health nurse plays a vital role in encouraging home care for MRs, giving parents guidance, and educating the community to the MR home care concept. (2 refs.) - *S. Glinsky.*

Binghamton City Hospital
Binghamton, New York

2302 BUCKINGHAM, DOROTHY. Wigs replace protective helmets. *Nursing Outlook,* 19(2):118-119, 1971.

Commercially available wigs provide protection to epileptic patients formerly protected with helmets. Three adult female epileptic patients in a state hospital have successfully used wigs for 10 months and during falls and accidents received protection equal to that provided by helmets. The synthetic wigs cost approximately $20 and are easily washed and maintained. In addition, the wigs have improved the patients' appearance and self-image, giving them a new pride in themselves. (No refs.) - *S. Glinsky.*

Porterville State Hospital
Porterville, California

2303 VON PREMERSTEIN, RICHARD. Zur Entstehung und Bedeutung des Begriffes 'Heilpadagogik' in der Anstalt Levana (The origin and significance of the concept 'pedagogic therapy' in the institution of Levana). *Heilpadagogische Forschung,* II(2):248-256, 1970.

In an effort to trace the origins of 'pedagogic therapy', the education of imbeciles in the 'Levana' institution is discussed. The term was first introduced by educators at this institution near Vienna in 1856. If therapy was to be successful, it was understood that medical treatment, mechanotherapy, and education must complement each other. 'Therapy' was conceived as the restructuring of the sick organism through a continuing, step by step process. Not only formal instruction, but also play and work were stressed. Eventually a special system of education was established specifically for idiots and difficult educable children. (9 refs.) - *P. Custers.*

Breslauer Str. 14
355 Marburg, Germany

2304 STEPHEN, ELSPETH; & ROBERTSON, JEAN. Normal child development and handicapped children. *Modern Perspectives in Child Psychiatry,* 1:210-234, 1971.

Handicapped children may be guided within a framework of normal child development. Systematic knowledge of perception, relationships, self-help, and intelligence, as each relates to normal development, can be applied to the handicapped. (53 refs.) - *E. L. Rowan.*

Foundation and Carshalton Hospital Group
Surrey, England

2305 PINE, NELLIE H.; & PERKINS, KAY. A successful "take" (after grafting an NIMH grant). *Perspectives in Psychiatric Care,* 9(1):8-12, 1971.

Nurses for the MR at the Patton State Hospital in California were successfully trained in behavior shaping techniques by personnel from the Pacific State Hospital where the program was being tried under an NIMH grant. The new program was found to result in improved behavior and capacity for enjoyment in the patients. (No refs.) - *E. L. Rowan.*

No address

2306 JONES, MARY. Artist communicates with retarded children. *Catholic Charities Review,* 55(2):16-19, 1971.

A retired artist teaches MR children to communicate by expression. The children are interested in learning to paint. Emphasis is on teaching the children to communicate anger, sadness, or pain by expressing themselves on paper, thereby ridding themselves of inner tensions. (No refs.) - *J. K. Wyatt.*

No address

2307 **BOBATH, BERTA; & FINNIE, NANCIE** R. Problems of communication between parents and staff in the treatment and management of children with cerebral palsy. *Developmental Medicine and Child Neurology*, 12(5):629-635, 1970.

Parental answers to a questionnaire on the home care and treatment of children with cerebral palsy revealed lack of understanding between parents and staff about the problems and needs of the children. The questionnaire was given to the parents of 45 children attending the Western Cerebral Palsy Centre, and 40 questionnaires were returned, 20 from parents of boys and 20 from parents of girls. The questionnaire sought information on the parents' opinions regarding the adequacy of training at the Centre, whether explanations about their children's problems were clear, and whether guidance of parents could be improved to obtain better cooperation. The results indicated that the use of technical medical terms should be avoided, that simple instructions regarding cerebral palsy and its attendant problems be given, and that parents should be informed in simple terms of the aims of treatment and of the Centre treatment program. Parents should also be kept fully advised of their child's progress. An improved atmosphere of cooperation and openness has subsequently developed, and a revised questionnaire is used as an integral part of the Centre's program. (1 ref.) - *S. Glinsky.*

Western Cerebral Palsy Centre
London, N.W.8., England

2308 **TURTON, LAWRENCE J.** Interdisciplinary panel presentation: Solving feeding problems of mentally retarded children—Speech pathologist. In: Springer, Ninfa Saturnino, ed. *Proceedings of a Conference on Nutrition and Mental Retardation.* Ann Arbor, Michigan, University of Michigan Publications Distribution Service, 1971, p. 100-106.

Biological activities of the cranio-oro-facial region, including feeding movements, do not necessarily facilitate speech development. Evidence suggests that the best way to improve speech is to teach a child to talk. Feeding sequences are necessary for health and well-being, but have little relevance to speech behavior. Also, the evidence for motor-oriented articulation development is open to question; articulatory defectives do not seem retarded in any specific general motor area. Cranial-oro-facial structures during speech and nonspeech activities do not appear related to clinical aspects of nonspeech exercises, including feeding. The speech-generating neuromuscular process seems uniquely organized and unrelated to processes underlying movements of the same muscle groups during other acts. It is possible to teach voluntary palatal movements even when S lacks awareness of movement. (18 refs.) - *B. Berman.*

2309 Orthomolecular medicine and the brain-handicapped child. *Expectations*, 1(1):2-3, 1971.

Research on the use of massive doses of vitamins and minerals in the treatment of brain-injured or neurologically handicapped children is reviewed. Nobel laureate Dr. Linus Pauling, who terms such treatment "orthomolecular therapy," contends that many mental dysfunctions may result from abnormal molecular concentrations of vitamins and other essential nutrients in the body. Correcting these imbalances may significantly improve the prognosis of mentally ill patients. Dr. Bernard Rimland, of the Institute for Child Behavior Research in San Diego, California, discounts the widespread belief that such vitamin therapy lacks scientific foundation, and claims that mentally ill children and adults do have markedly atypical vitamin metabolism. Research by Dr. Alan Cott, of the New York Institute for Child Development, indicates that positive results are more than mere placebo effect; when megavitamin therapy is interrupted, symptoms reappear. In all cases, substance type and dosage are dependent on individual need. Side effects of large doses of niacin, which is frequently used in such therapy, are being investigated, and its efficacy in treating some types of schizophrenia is questioned. (12 refs.) - *M. H. Street.*

PROGRAMMATIC ASPECTS – Planning and Legislative

2310 U. S. HEALTH, EDUCATION, AND WELFARE DEPARTMENT. Developmental Disabilities Services and Facilities Construction Act – Public Law 91-517. *Programs for the Handicapped*, 70-5, December 7, 1970, 9 p.

The provisions of the 1970 Developmental Disabilities Services and Facilities Construction Act (PL 91-517) significantly expand the scope and purposes of the amended MR Facilities Act of 1963. The provisions of PL 91-517 include services for all MRs, as well as for those with developmental disabilities originating in childhood. The states have a broad responsibility for program planning and implementation. Local communities will be involved in specifying needs, establishing priorities, and actual delivery of services. The new law includes a combined formula and project grants program which covers the construction of facilities and the provision of services. Present authority to construct university-affiliated facilities for the MR has been extended through June 30, 1973, and a new program covers the costs involved in the administration and operation of demonstration facilities and their interdisciplinary training programs. The individual states must submit state plans which fulfill specifically detailed requirements before they can qualify for federal assistance under a new formula grant program. A National Advisory Council on Services and Facilities for the Developmentally Disabled will advise the Secretary of Health, Education, and Welfare on regulations and will evaluate programs. Federal appropriations for this program are $60 million in fiscal year 1971, $105 million in fiscal year 1972, and $130 million in fiscal year 1973. (No refs.) - *J. K. Wyatt.*

2311 *Mental Retardation Financial Assistance Programs of the Department of Health, Education, and Welfare.* Superintendent of Documents, U. S. Government Printing Office, Wash., D.C., 1971, 70 p.

This guide to the Department of Health, Education, and Welfare's MR programs – which range from community day - care center support to the construction of a complex diagnosis and evaluation facility – summarizes the programs' financial-assistance aspects and attests to the Nation's ever-increasing commitment to solving the problems of the retarded. Produced with the cooperation of the President's Committee on MR, the booklet covers 5 major program categories – Basic and Supportive Services, Research and Demonstrations, Construction, Training, and Income Maintenance – and a miscellaneous group of programs such as the Foster Grandparent Program, Head Start, and Surplus Property Utilization for Education and Public Health Purposes. Information on each program includes: purpose; who may receive funds; the office that provides additional information; and references to authorizing legislation. Three types of grants are discussed: formula, project, and individual. A list of regional offices is also included. (No refs.) - *B. Berman.*

2312 *U.S. Mental Retardation Construction Program.* Health, Education, and Welfare Department, Washington, D.C., 1971, 99 p.

The Developmental Disabilities Services and Facilities Act of 1970, which amends the Mental Retardation Facilities and Mental Health Centers Construction Act of 1963, provides Federal support for construction of facilities designed for persons with mental retardation and other developmental disabilities. Research centers to study causes and means of prevention of MR, university-affiliated facilities for the training of personnel for research and service careers, and community facilities for diagnosis, treatment, education, training, care, and sheltered workshops are also discussed. (No refs.) - *D. Fergerson.*

2313 MARSHALL, TONY. Voices for the silent minority. *Mental Health*, Autumn: 40-42, 1970.

The handicapped must often rely upon spokesmen to protect their rights, since they are frequently unable to do so themselves. During the past few years efforts have increased on behalf of the handicapped. The lobby of the National Society for Mentally Handicapped Children, along with other groups, has ensured that the handicapped child has the same right to education as the normal child. The Mental Health Act of 1959, which conferred the right of informal admission to the hospital for the mentally handicapped, constituted an earlier milestone. At the present time the handicapped are also protected against ill-treatment and sexual exploitation, and they have the right to vote and marry. Within the hospital, the key figure is the charge nurse, who sets the tone and the pace of a ward. The National Association of Mental Health, health service trade unions, and professional organizations are presently trying to establish a code of practice for nurses. (No refs.) - *B. J. Grylack.*

No address

2314 **SIMPSON, P. F.** Facing the new challenge. *Special Education*, 59(4):15-17, 1970.

Transfer of responsibility for the control of junior training centers from local health authorities to local education authorities should reduce the great variations in control over the centers. Because of the prevalence of untrained staff in many centers, most heads do not have or do not believe they have the independence experienced by the heads of schools under local education authorities. Restrictions can be imposed by the local health authorities because legal precedents regarding the responsibilities of teachers in training centers have not been established. Educators must control educational principles and methods used in junior training centers. Inter-staff relations will need to improve to ensure cooperation between experienced teachers and those who are newly trained. Teachers will need to reappraise their aims and methods and develop necessary attributes as well as realistic expectations. (No refs.) - *J. K. Wyatt.*

Newcastle University Institute of Education
Newcastle-upon-Tyne, England

2315 **CLAUSEN, JOHS.** The plight of special education. *Journal of Special Education*, 4(2):157-159, 1970.

Although increased funds for research in the education and habilitation of MRs are needed, Albee should not promote educational research at the expense of research in other disciplines. Shifts in research priorities from biomedical to special education should be based on the merits of research proposals rather than on bureaucratic decree. The improvement of educational procedures and facilities for MRs is justified by humanitarian and social arguments and is not related to the issue of etiology. Since MRs have never received the special educational helps they need, it is not possible to predict, as Albee does, that 85% will benefit from these programs. The solution to the need for research funds in special education lies in the development of a series of specific and constructive research proposals aimed at establishing the effects of specific educational procedures on the behavior of MRs. (5 refs.) - *J. K. Wyatt.*

New York State Institute for Basic
Research in Mental Retardation
Staten Island, New York

2316 **CALLANDER, D. J.** As normal a life as possible is the aim in Sweden. *Intellectually Handicapped Child*, 9(4):24-25, 1970.

The education, training, and care of MRs in Sweden are based on "normalization," a philosophy which mandates that all facilities available to normal children and adults be made available also to MRs. Sweden has expanded greatly its facilities for special education and training, and the trend is toward increasingly smaller classes and institutions. The emphasis is also on integrating the handicapped into the community and providing services for the MR. The gradual relinquishing of private facilities to the state has the disadvantage of diminishing parental interest and involvement, which has implications for the Intellectually Handicapped Children's Society in New Zealand. The Mockasinen Day School in Stockholm for EMRs, a showpiece facility, houses EMRs in 6 villas and has an impressive educational program and record. (No refs.) - *S. Glinsky.*

No address

2317 Much more data needed on intellectual handicap. *Intellectually Handicapped Child*, 9(4):4, 1970.

The Intellectually Handicapped Children's Society must more vigorously gather reliable, quantitative data on intellectual handicap in New Zealand. A preliminary survey of the Society's records reveals inadequacies in the case records system which preclude accurate assessment of the Society's condition, accomplishments, or programs. Reliable, complete case records must be initiated to increase professionalism, facilitate local planning needs, develop programs on a national level, and enable the Society to justify continued support from the government, organizations, and individuals. (No refs.) - *S. Glinsky.*

2318 MADELEY, B. R. A host of new ideas. *Nursing Mirror,* 132(3):28-29, 1971.

The number of institutionalized MRs should be reduced to include only those who need hospital care. MRs should receive treatment for their various handicaps and their condition should be diagnosed and treated specifically, not in generic terms. Placing MRs in small homes of 6 to 8 rooms would provide a more domesticated and natural environment. In order to stimulate imagination and enthusiasm in hospital staffs, refresher training courses that reflect the movement away from custodial care toward rehabilitation, social training, and education may be needed. Programs have been developed in which nurses help MRs shop for clothing and develop recreational plans. (No refs.) - *J. K. Wyatt.*

Mid-Sussex Hospital
Sussex, England

2319 The mentally handicapped today. *Nursing Mirror,* 132(2):10-11, 1971.

A conference was held to create a greater understanding of problems involved in the care of MRs and to establish closer relationships between statutory and voluntary organizations. Papers delivered included information on the importance of the pediatrician, educational provisions for SMRs, problems of national policy related to SMRs, and the need for additional community services. The needs of a family with a handicapped child were discussed by a parent. (No refs.) - *J. K. Wyatt.*

2320 U. S. HEALTH, EDUCATION, AND WELFARE DEPARTMENT. *Programs for the Handicapped,* 71(3):1-15, 1971.

Federal funds for learning disabilities programs and projects are provided for by Section G, Title VII, of Public Law 91-230. Funds are administered by 3 major divisions of the Bureau of Education for the Handicapped: Educational Services, Training Programs, and Research. New approaches to funding are under consideration. Model centers will provide evaluation services, develop model programs, assist other agencies in the development of model programs, and disseminate new methods and techniques. Financial support is available to expand professional training in the area of learning disabilities. Support has been provided for research projects related to assessment, diagnosis, remediation, and/or classroom instructional arrangements. The National Institute of Neurological Diseases and Stroke supports research in education, medicine, and psychology. Although most of the funds provided by the National Institute of Child Health and Human Development are for studies of normal learning processes, some projects are concerned with learning disorders or with the relationship between learning and MR, or physical or cultural deprivation. Rehabilitation Services Administration is involved in learning disabilities in a regular State-Federal program, a new evaluation and work adjustment program, and a new program in developmental disabilities. The Office of Education will support demonstration projects related to health and nutrition. (No refs.) - *J. K. Wyatt.*

Secretary's Committee on Mental Retardation
Washington, D.C. 20201

2321 Retardation: An environmental problem. *Performance,* 21(7):9-10, 1971.

Many findings revealed by current research are finally indicating that it may be possible to reduce the devastation produced by the age-old problem of MR. A study conducted by the National Association for Retarded Children emphasizes that any lack of knowledge about MR is matched by information on certain predictable patterns associated with the environment of poverty. Since voluntary and professional agencies have worked primarily with MR in the middle class and have not reached the real center of congenital and environmental retardation in the poverty areas of the country, the Association is seeking a reevaluation of this pattern. If the need for serving the entire community and for attacking the problem of inadequate health care and other retardation-causing factors is met, the

number of needlessly retarded children in impoverished areas of the United States can probably be reduced. (No refs.) - *B. J. Grylack.*

2322 **JERVIS, GEORGE A.** Toward a brighter day for retardates. *Medical World News,* 12(2):44D, 1971.

Former neglect of MRs has given way to deep concern for MR, and a comprehensive MR care and management program must be developed and articulated. Although traditional definitions of MR stress intellectual deficiency, increasing importance is being attached to social and behavioral adjustment of the MR. A comprehensive MR care program would include identification and diagnosis, medical care, home care, management in schools, vocational rehabilitation, and residential care. Such a broad-based program unquestionably improves the performance, if not the intelligence, of the MR, prevents mental deterioration, improves habits and the socialization process, and often normalizes emotional imbalance. (No refs.) - *S. Glinsky.*

Columbia-Presbyterian Medical Center
New York, New York

2323 **KARCZEWSKI, MIECZYSLAW.** L'aide sociale aux debiles mentaux en Pologne (Social assistance to the mentally retarded in Poland). *Nos Enfants Inadaptes,* 36:18-22, 1970.

The Polish system of social services can be defined as a system aimed at complete analysis of the social needs which exist and at a constant increase in the efforts for their maximal satisfaction, through the most rational use of the available means. The role of the state is crucial. Two ministries share primary responsibilities in the field of social action: the Ministry of Health and the Ministry of Education. The responsibility for care of MR children falls only partly under the jurisdiction of these governmental agencies. Presently there are 200 institutions for the mentally retarded, of which 110 are designed specifically for children. Re-education (which includes medical, social and professional aspects) is the highest task of these institutions. Every retarded child is considered re-educable and an optimistic view is taken of the future prospects for care of the MR, considering the possibilities for continued progress in technology and the human sciences. (No refs.) - *P. Custers.*

No address

2324 **NIRJE, BENGTO.** Towards independence. *Deficience Mentale/Mental Retardation,* 21(2):2-8, 1971.

At the 1969 Congress of the International Society for Rehabilitation of the Disabled (Dublin, Ireland) a description of the continuing efforts being made in Sweden to attain more respect and independence for the MR was presented. Institutions and services for the retarded are changing, as are prejudices and public attitudes. Three interdependent but not equally curable handicaps, the 'burdens of the retarded', are distinguished: the mental retardation of the individual; the imposed or acquired retardation; and the awareness of being handicapped. The Normalization Principle, upon which Swedish MR programs are based, aims at ameliorating these burdens for the entire developmental range of MR by making available patterns and conditions of everyday life which are as close to normal life experiences as possible. In this respect, some positive experiences of the strengthening opportunity afforded by clubs for the adult retarded are described. At conferences of retarded adults, the initiative of the retarded themselves in discussing views and ideas of their own concern has significantly improved general understanding of the problems of retardation. (No refs.) - *P. Custers.*

No address

2325 Toekenning van de 'gewone tegenmoetkoming' aan sommige zwaar verstandelijk gehandicapten ('Normal support' for some cases of seriously mentally retarded children). *Amentia,* 23:12-13, 1971.

Belgian law distinguishes two kinds of financial support for retarded children: 'normal support' (amounting to maximal 50,000 F per year) and 'special support' (around 36,000 F per year). The first category is intended for physically retarded children; the second one, in general, for the mentally retarded. The aim of the National Association for Aid to the Mentally Handicapped is

to bring the support for mentally retarded up to the level of that for the physically retarded. Specific cases in which the mentally retarded can, on the basis of the existing law, claim 'normal support' are discussed. (No refs.) - *P. Custers.*

2326 10,000 arrieres profonds en Belgique: Que peut-on faire pour eux? (10,000 seriously retarded in Belgium: What can one do for them?). *Amentia,* 23:8-11, 1971.

Special attention should be given to the problems of the profoundly MR in Belgium, particularly in those cases where physical handicaps accompany the mental impairment. There must be a special effort to alleviate the everyday problems faced by the families of these MR children who elect to keep the child at home. In addition to the services which are already available for the MR, there is a need for special kinds of aid aimed directly at this group. Day care centers, medical and educational clinics, and facilities for the adult MR need to be constructed. Access to babysitters and family servants should be made available to all affected families, with government financial aid forthcoming where necessary. Additionally, budgetary support from the government should be greatly increased. (No refs.) - *P. Custers.*

2327 A look into the future for systems of retardation services. *Deficience Mentale/ Mental Retardation,* 21(1):2-8, 1971.

In a recent address to a Canadian conference on MR, an American expert stressed the need for strong citizen support for future guarantees of the rights of all citizens, including retardates. Planned comprehensive services must replace crisis-oriented programs, and extensive diffusion of specialty services into generic services, especially through the medium of public and early childhood education, is a necessary change. It is predicted that there will be objectification of human management with universal acceptance of decision-making theory, extensive consumer participation, and new approaches to residential services in the future. (No refs.) - *B. J. Grylack.*

2328 Education of mentally handicapped children. *Lancet,* 2(7675):730-731, 1970.

The recently adopted English Education (Handicapped Children) Act of 1970 transferred responsibility for the education of handicapped children to the local education authorities. A child previously considered unsuitable for education at school will be classified as requiring special educational treatment. This will permit adoption of facilities to the individual needs of each child. Existing buildings will be transferred to local educational authorities. An interim one-year reorientation course for the present teachers will suffice pending inauguration of a regular 3-year program for teachers of the mentally handicapped. (No refs.) - *E. Kravitz.*

2329 Bridging the gap. *Medical Insight,* 3(1): 45, 1971.

The Task Force on the Mentally Handicapped, in a 4-month review of the status of the nation's 25 million MRs and mentally ill, urges expansion, through federal legislation, of community services for retardates and a continuing shift away from institutional care. (No refs.) - *B. Berman.*

2330 ALMACK, RONALD B.; & DiGIOVANNA, FRANK. Developmental Disabilities Services and Facilities Construction Act. *Rehabilitation Record,* 12(2):23-25, 1971.

On October 30, 1970, the Developmental Disabilities Services and Facilities Construction Act became law, amending the Mental Retardation Facilities and Community Mental Health Centers Construction Act, the basic MR authority. In the Act developmental disability is defined as a disability attributable to MR, cerebral palsy, epilepsy, or another neurological condition of an individual requiring treatment similar to MR treatment. The Act authorizes appropriations for formula grants to states, grants for projects of national significance, grants for interdisciplinary training programs in institutions of higher learning, and grants for university-affiliated facilities. It will be administered by the Rehabilitation Services Administration. (No refs.) - *S. Glinsky.*

No address

2331 WATTS, ANNE P. The developmental disabilities act. *Journal of the American*

Speech and Hearing Association, 13(7):391-393, 1971.

The Developmental Disabilities Services and Facilities Construction Amendments of 1970 became public law 91-517 in October of 1970, expanding the scope and purposes of the Mental Retardation Facilities Construction Act of 1963. The new law is designed to provide the states with broad responsibility for planning and implementing comprehensive service programs for all MRs or those with other developmental disabilities. It also gives communities a decisive voice in determining needs, establishing priorities, and developing vehicles for delivery of services to the MR and developmentally disabled. (No refs.) - *S. Glinsky.*

American Speech and Hearing Association
Washington, D. C. 20014

2332 **GARRETT, JAMES F.; & GRIFFIS, BERTRAM W.** The economic benefits of rehabilitation for the mentally retarded. *Welfare in Review*, 9(2):1-7, 1971.

Both patients and society benefit from rehabilitation programs for MRs. A review of rehabilitation programs in terms of costs and statistics supports such investments. Both federal and state training and placement programs show returns on funds spent on program research and administration. Of more importance than the proven economic gains are the human benefits and decreasing prejudice towards MRs. (9 refs.) - *S. Glinsky.*

No address

2333 **ALBEE, GEORGE W.** In reply (Emerging conceptions of mental illness and models of treatment). *Professional Psychology*, 2(2):142-144, 1971.

Alternate suggestions are made in reply to Drs. Grinker, Garmezy, and Schachter's ideas regarding emerging conceptions of mental illness and models of treatment. The discussion was held at the American Psychological Association and American Psychiatric Association meetings. The illness model could be simplified to permit more people to be treated and make more interventions available. In addition, tax funds should be spent on education and rehabilitation instead of institutions for the retarded, criminal and delinquent, until biological discoveries facilitating prevention and alleviation are made. (No refs.) - *S. Glinsky.*

No address

PROGRAMMATIC ASPECTS — Community

2334 **STONE, NELLIE D.** Effecting interdisciplinary coordination in clinical services to the mentally retarded. *American Journal of Orthopsychiatry*, 40(5):835-840, 1970.

Clear definition of roles and division of labor are necessary to provide clinical help to the retarded in schooling, health care, socialization, vocational training, and interpersonal relationships. Skills in being independent, interdependent, rebellious and in being able to resolve conflict in other than win/lose ways should be encouraged. Techniques have been implemented to enable the clinician to communicate freely, share responsibility, respect individual status, encourage independent and interdependent functioning, develop congenial interpersonal feelings and evaluate clinical functioning. (9 refs.) - *G. S. Bell.*

Newark State College
Union, New Jersey 07083

2335 **FORREST, D. M.; BOWEN, TREVOR; WALKER, GEOFFREY F.; & SIMPSON, ELIZABETH D. L.** Coordinated care of the child with spina bifida. *Lancet*, 2(7679):934-935, 1970. (Letter)

Experience with more than 700 children at Queen Mary's Hospital for Children indicates that combined clinics are neither necessary nor helpful when such large numbers of spina bifida patients are involved. A satisfactory arrangement has been worked out where separate clinics in 3 medical specialties – pediatrics, orthopedics, and ophthalmology – handle the vast majority of spina bifida cases. Ancillary personnel are freely available when needed. (No refs.) - *G. S. Bell.*

Queen Mary's Hospital for Children
Carshalton, Surrey, England

2336 **DAVIDSON, V. TRENT.** A new day for Sam and Abby. *Spectrum*, 45(6):4-6, 31-33, 1969.

A growing public awareness of responsibility for the MR is producing new methods, materials, and skills, and a knowledge that MRs should be treated, as much as possible, as normals. Churchmen, responding to social pressures, are instructing MRs in the nature of God, love of Jesus Christ, and revelation of the love, hope, and community orientation of Christianity. The church is producing suitable materials for both educable and trainable retardates. Materials for younger educables focus on self-worth and acceptance, and are designed for the volunteer teacher. Churches are organizing special committees that cooperate with community groups in establishing priorities, developing plans for leadership training, providing direct-involvement experiences, and furnishing material resources. (No refs.) - *B. Berman.*

No address

2337 The Foster Grandparent Program. A report – 1970. *Programs for the Handicapped*, 70(2):2-3, 1970.

The Foster Grandparent Program – developed by the Department of Health, Education, and Welfare in 1965, funded by the Office of Economic Opportunity, and formally established by Congress in 1969 – has proved beneficial to both children and the elderly. Retarded children in this program have shown improvement in self-care and motor skills; spastic children have learned to feed themselves. The foster grandparents have shown increased self-esteem, a sense of purpose, improved health, and greater financial security. They have benefitted also by counseling and opportunities to associate with members of their own generation. From an initial 21 projects the program has expanded to 68 in 40 states and Puerto Rico, with more than 4,000 persons aged 60 or over working with children in over 180 settings, 4 hours a day, 5 days a week. The program's potential for further expansion remains undiminished. (No refs.) - *B. Berman.*

U. S. Department of Health, Education and Welfare
Washington, D. C. 20201

2338 **BROWN, BERTRAM S.; & COURTLESS, THOMAS F.** *The Mentally Retarded Offender.* DHEW Publication No. (HSM) 72-9039. Superintendent of Documents, U. S. Government Printing Office, Washington, D. C., 62 p.

This survey of the problems pertaining to the MR offender stresses that understanding the offender yields new insights into the whole complex of retardation and the area of anti-social behavior generally. An introductory historical review of the 3 periods of professional and societal interest in this problem (Early Enthusiasm: 1890-1920; Denial and Neglect: 1921-1960; The Contemporary Scene: 1961-present) reveals the changing policy emphases from viewing the MR offender as almost always feeble-minded, to a later relegation of retardation in criminal behavior to a secondary, almost ignored, position, to the recent strong interest in the association between retardation and anti-social behavior. The survey covers: Management and Treatment of Retarded Offenders; Defective Delinquent – Changing Definitions; The Institutionalized MR Offender (Major Survey, Follow-Up Survey of Inmates with IQs <55, Management Problems and Practices, Case Study of an Inmate Sample Analyzed for Socio-Psychological and Legal Variables); Current State Planning for Retarded Offenders; and Critical Issues and Recommendations for Action in Research. (No refs.) - *B. Berman.*

National Institute of Mental Health
Rockville, Maryland 20852

2339 **HUNT, J. McV.** Parent and child centers: Their basis in the behavioral and educational sciences. *American Journal of Orthopsychiatry*, 41(1):13-38, 1971.

Parent and child centers constitute a limited deployment of an innovative attempt to intervene constructively in the educational functioning of poor families and neighborhoods. Intervention, the primary concept in these centers, is based upon the beliefs that plasticity of behavioral development exists, that there is a mutual interrelationship between poverty and the lack of competence, that competence does not develop automatically in genotypes of even the highest potential, and that poor parents can be taught to be effective teachers of their young children. Whereas attempts to modify the child rearing practices of parents through professional talk alone have failed completely, poor parents can learn to improve child rearing methods through opportunities to witness the manner in which relatively skilled teachers or social workers manage children either in nursery school or in home demonstrations, especially when these demonstrations are explained in comprehensible language and when the mothers are respected. (94 refs.) - *B. J. Grylack.*

University of Illinois
Champaign, Illinois 61820

2340 **WILLIAMS, NORWOOD L.; SABATINO, DAVID A.; & KESMODEL, KRISTIN.** A community program directed at breaking the poverty-retardation cycle. *Journal of Rehabilitation*, 36(4):35-36, 1970.

It is generally difficult to distinguish clearly between the external manifestations of cultural influences and those inherent problems related to MR, especially among inner city populations. In an attempt to interrupt the poverty-MR cycle for ghetto residents, the Industrial Sewing School was established in Baltimore in 1960 to train middle-aged adults in the vicinity, most of whom were unemployed welfare recipients or part-time maids who functioned within the mild MR range of intelligence and had a mean IQ of 74. In 1967 a Satellite Rehabilitation Program was opened at the school to provide services and training. The majority of the clients of the school receive support from the Division of Vocational Rehabilitation. Counseling interviews are held weekly for new applicants at the school, and weekly psychological assessment is also provided. After the diagnostic services are provided and a client is declared eligible for the services of the agency, she is enrolled in the 4-month intensive training program. Most clients have completed the course successfully and have maintained gainful employment for a minimum of 2 months. They are considered rehabilitated, and most important, their human dignity has been reestablished. (No refs.) - *B. J. Grylack.*

No address

2341 **JEPSON, ANNE M.** Coordinated care of the child with spina bifida. *Lancet*, 2(7683):1140-1141, 1970. (Letter)

Local-authority facilities can be very helpful in the management of children with spina bifida. The medical assessment unit of the school for handicapped children sees children at an early age; it serves 6 London boroughs with diagnostic and therapeutic facilities. Liaison is maintained with each child's hospital and between the teaching staff and unit personnel. Learning difficulties are considered. Unfortunately, there is still too little concern for the adult life of these handicapped children. (2 refs.) - *E. Kravitz.*

Medical Advisory Unit
Hounslow, Middlesex, England

2342 Center for low IQ's. *Rehabilitation Record*, 11(6):7-9, 1970.

The Cooperative School Rehabilitation Center at Minnetonka, Minnesota, provides a comprehensive developmental program for young people, ages 14 to 21, whose low IQs (all below 60, some between 15-30) prevent their adapting to public school. Each student maintains a continuing relationship with a case manager, while progressing through a manipulation laboratory and a series of personal and social development activities. Prevocational training and practical work experience are mingled with innovative activities that include a food service program, Montessori teaching methods, music instruction, transportation training, and physical education. New methods and ideas are applied in teaching situations with emphasis on reality. Employment is

the ultimate goal: of 91 who had left the center by January 1969, 55 are working full time in competitive employment. (No refs.) - *B. Berman.*

2343 **CORTAZZO, ARNOLD D.; & RUNNELS, EUGENE J.** One approach in rehabilitating the retarded. *Rehabilitation Literature,* 31(12):354-360, 1970.

The principal objective in rehabilitation of the retarded is to render them as capable as possible of meeting their adult responsibilities. A training center has devised a new rehabilitation plan which is based upon 4 program divisions, including vocational rehabilitation, education and training, independent living, and development and training. Through this plan, the retarded are trained and placed in appropriate employment, are equipped with social and community skills, and are placed in appropriate community living situations. The program utilizes vocational rehabilitation counselors, psychologists, social workers, physicians, nurses, recreation therapists, training instructors, and placement counselors to achieve its goals. Facilities are geared as much as possible to the living and working needs of clients. (8 refs.) - *B. J. Grylack.*

Sunland Training Center
Miami, Florida

2344 **WALLACE, HELEN M.** How to evaluate the services and care offered handicapped children. *Public Health Reports,* 85(7):647-655, 1970.

Evaluation can be used to analyze the care received by some handicapped children and to identify unmet needs. Factors to be considered in the evaluation of casefinding include: age at which selected types of handicapped children are identified; age of onset compared to age at identification; use and accuracy of screening procedures; use of birth certificates in early casefinding; and comparison of known handicapped children with incidence and prevalence information. Evaluation of diagnostic, treatment, and rehabilitation services should include demographic aspects of care, results, caseloads, care standards, and continuity of care. Other areas which should be carefully evaluated are educational and vocational services; day, foster home, and residential care; and transportation services. The effectiveness of prevention programs, agency relationships, and barriers to the use of services and resources should also be carefully evaluated. (10 refs.) - *J. K. Wyatt.*

University of California
School of Public Health
Berkeley, California

2345 **O'CONNOR, GAIL; JUSTICE, R. S.; & WARREN, NEIL.** The aged mentally retarded: Institution or community care? *American Journal of Mental Deficiency,* 75(3):354-360, 1970.

A majority of an analyzed group of institutionalized, aged MRs was judged eligible for community placement. The Ss were 463 MRs over 60 years of age who were considered for community placement by ward personnel in 19 state institutions. Fifty-nine percent of the group was judged eligible for community placement on the basis of higher intelligence, passivity, few physical handicaps or behavior problems, and the ability to do work tasks. Later analysis added 69 persons, bringing the total eligible for placement to 74%. The fact that these MRs were not placed seems related to lack of effective out-of-hospital leave programs, the MRs' usefulness as ward helpers, and hospital personnel's doubts regarding better care being given in a community setting. Yet such placement is preferable for economic and humane reasons. (21 refs.) - *S. Glinsky.*

Pacific State Hospital
Pomona, California 91768

2346 **ZEROLIS, JOANNE B.** Preparing long-term retarded residents for community placement. *Hospital and Community Psychiatry,* 72(5):32-34, 1971.

Work with 2 groups of long-term retarded hospital residents (5 women, average IQ, 38; 5 men, average IQ, 41) demonstrated surprising behavior changes preparatory to community placement. Working with these passive-dependent Ss required overcoming their resistance and that of the staff. Reaching out to these old-timers, showing concern for each in various situations and thus emphasizing his unique importance, proved the

most effective way of changing behavior. This approach was reinforced by group experiences that captured the Ss' interest as well as physical presence. It was also important to allay fears of failure—of being exposed as incompetent—and to help develop communication skills. Behavior cues and gestures, as well as group decision-making, such as planning community trips, role-playing, or sociodrama, were effective in this respect. An important need that developed was the requirement to measure time and to gain perspective. Home touring proved very successful in clarifying the meaning of community placement; it reflected a need to bring the community and the old-timers together in mutual understanding. (No refs.) - *B. Berman.*

The Oaks-Hospitals and Health Centers
San Jose, California

2347 Handicapped children. *British Medical Journal*, 3(5716):179, 1970.

Living with Handicap, a report published by the National Bureau for Co-operation in Child Care, is based upon data compiled by a multidisciplinary working party. Although the information gained from solicited letters from 412 parents is biased, since it represents the views of the more articulate, intelligent, and socially conscious segment of the community, the replies are poignant and emphasize the feeling of hopelessness engendered by the rigid departmentalization of services available to the handicapped. The report is concerned with the problems of families trying to cope with children with physical and mental handicaps, and it reveals that the general public tolerates physical handicap much more easily than mental handicap. The ultimate plea of the working party is that each child should receive help according to individual needs and that this assistance should be provided with minimum separation from and disturbance of normal family life. (5 refs.) - *B. J. Grylack.*

2348 NAYLOR, HARRIET. New trends in volunteer services for the mentally handicapped. *Hospital & Community Psychiatry*, 22(4):109-111, 1971.

As volunteer work is becoming increasingly popular, it is stimulated in the mental health field particularly by emphasis on community treatment for the mentally ill and retarded. Caretaking, social sustenance, professional assistance, developmental rehabilitation, and advocacy are among the functions which have been suggested for volunteers in this field. Since the handicapped and retarded have difficulty lobbying for their own interests, volunteers may be valuable assets in helping to direct community resources to the problem. Volunteers benefit from the services they provide by developing their own skills. Although regular paid professional staffs may be reluctant to accept the volunteers initially, careful organization and an increase in the confidence of the volunteers themselves will lead to an effective working relationship and to a new dimension in care for the handicapped. (1 ref.) - *B. J. Grylack.*

State Department of Mental Hygiene
Albany, New York

2349 BROWN, RICHARD J. Brain damaged adolescents: Their miseducation in a rehabilitation center. *American Journal of Orthopsychiatry*, 42(2):326-327, 1972.

A prevocational-educational-therapeutic program for brain injured adolescents, jointly written by a major public school system and a voluntary rehabilitation agency, resulted in multiple instances of child abuse. The adolescents were engaged in activities such as cleaning toilets, cleaning cafeteria tables, and packing denture cups 15 hours per week. Attempts to rectify the situation and upgrade the vocational activities have been fruitless. Despite the agency's justifications and professed realistic attitudes, the chief aim of the program is the self-perpetuation and self-aggrandizement of a program-depleted and client-hungry agency. No serious effort is being made to improve the program for the brain damaged adolescents. (No refs.) - *S. Glinsky.*

New York School of Psychiatry
Ward's Island, New York

2350 KAHN, MAX. The Hebrew Academy for Special Children. *Jewish Parent*, 22(3):32-34, 1971.

The Hebrew Academy for Special Children differs from the usual Jewish community educational center in that its special population represents a cross section of handicapped children from Jewish communities in the New York area. The overall approach to teaching is based upon interdisciplinary diagnosis, psychoeducational learning assessments, and performance surveys taken periodically of each student. Instruction in appropriate skills is delivered with the aid of various visual, motoric, kinesthetic, and auditory training techniques. Parental and public education on the problems and needs of the handicapped child is a crucial element in the program. The curriculum is geared towards success at various levels: some children will return to normal educational programs, some will participate in work-study programs and will be able to compete for a paid job, and others will require a sheltered workshop setting with continued supervision. (No refs.) - *B. J. Grylack.*

Hebrew Academy for Special Children
Brooklyn, New York

2351 **SOPER, HARRY H.** Community services of the Ontario hospital school, Orillia. *Deficience Mentale/Mental Retardation,* 21(2):12-14, 1971.

The O.H.S. Orillia Community Services Organization offers an alternative to traditional models of mental health clinics. Instead of approaching the problems of retarded children in isolation, these Community Services consider the MR child as part of a living unit—most immediately represented by his family and extensions into the community. Since the public health organizations were already dealing with the needs and problems of children, integration into these institutions of diagnosis and counseling services for parents of children who were having problems of an intellectual-emotional nature was seen as the natural and desirable alternative. Some positive impressions of work with community-based personnel and indications of beneficial effects of this program with respect to family adjustment are described. (No refs.) - *P. Custers.*

Ontario Hospital School, Orillia
Ontario, Canada

2352 **TANNER, JOHN.** A community oriented group home program for the mentally retarded. *Deficience Mentale/Mental Retardation,* 21(2):23-29, 1971.

The group home program in New Brunswick, Canada, developed as one element in the Regional Demonstration project in Mental Retardation, one of 14 projects sponsored by the Canadian Association for the Mentally Retarded. The purpose of the project is to develop coordinated services for the mentally retarded of all ages within a population area of 71,000 urban dwellers and 62,000 rural dwellers. The concept behind the project changed from the provision of a small institution with special services to the eventual establishment of a number of separate houses providing a family atmosphere and using regular services available to the community as a whole. Through the supporting educational and occupational programs in the community and the involvement of professional and business personnel, many training problems and financial costs could be successfully met. Two objectives, specifically, are attained: a changing of community attitudes towards the problem of MR; and the provision of a comfortable and satisfying living environment, leading to the maximum possible integration of the MR into regular community life. (No refs.) - *P. Custers.*

No address

2353 **POSTERNAK, YVONNE.** La participation des parents et des volontaires dans nos institutions (suite et fin). [The participation of parents and volunteers in the work of our institutions (follow-up and conclusions)]. *Nos Enfants Inadaptes,* 36:16-17, 1970.

Since many different kinds of social organizations currently provide volunteers for MR institutions, some standards have to be established for the quality of these volunteers. Generally, they should exhibit respect for the mentally retarded, a willingness to understand their problems, a sense of responsibility, and personal discipline. From the other side, institutions should provide adequate training for the prospective volunteers and should contribute a certain amount of social benefits. Efforts should also be made to improve staff-volunteer relationships with the particular role of each group clearly defined in advance, so as to forestall problems. (No refs.) - *P. Custers.*

No address

2354 **HARSHMAN, FRED.** Report on the Regional Mental Retardation Program in Saskatchewan. *Deficience Mentale/Mental Retardation*, 21(3):22-26, 1971.

The Regional Mental Retardation Program in Saskatchewan operates on the premise that the retarded individual, his family, and his community constitute an interacting and indivisible unit which should be maintained as such. One of the primary goals of the program is the prevention of institutionalization where possible. Community-based staff are considered necessary for the adequate functioning of community services, and it is felt that decentralized services are vital within the community approach to MR. One of the facets of this new approach is a community training home where children may be sent for a toilet or feeding problem, which is dealt with directly during a short-term training period in a homelike setting. The program has succeeded in implementing planned problem solving instead of crisis reaction, in improving the utilization of existing facilities, and in developing new resources. (No refs.) - *B. J. Grylack.*

Ontario Department of Health
Ontario, Canada

2355 Who can be a citizen advocate? *Deficience Mentale/Mental Retardation*, 21(3):16-18, 1971.

Citizen advocates are needed for impaired children without friends or relatives, impaired patients in nursing homes, impaired children living with impaired parents, impaired adults living independently without access to guidance, and impaired young adults living in hostels who require community adjustment. An advocate must be a contributing, stable, and competent member of the community who is committed to the concept of advocacy and who is willing to undergo orientation and preparation for the advocacy task and to join relevant community action groups. Residents of institutions are returning to their home communities under the guidance of their citizen advocates, who are learning about MR and working to reduce the injustices of the past. (No refs.) - *B. J. Grylack.*

2356 **deBABRA, MAREIA.** A junior training school in Ecuador. *Teaching and Training*, 9(1):25-27, 1971.

The "Centro Psico-Pedagogico," the first school for retarded children in Ecuador, was established in 1966 as a day school for 18 EMRs of CA 5 to 12 with a full-time staff of 4. Fees were assessed according to each family's ability to pay. After 2 years the school became a foundation and as such now receives about half its financial support from the Government. The staff has increased considerably, and the 42 children receive their instruction in groups of 10 graded according to CA (except for one group of TMRs). The program consists mainly of communication skills, motor skills, self-help, arts and crafts, and visual perception. Parents' reactions to the school are mixed: they are grateful for what is being done for their children but also are forced to accept the condition of retardation. Approximately 60 - 70% of the parents attend the quarterly meetings organized for them. Although there are now 2 other small groups for MRs in Guayaquil, there is no possibility of further training for the young unless a workshop for the older boys and girls can be organized. (No refs.) - *J. C. Moody.*

Centro Psico-Pedagogico
Guayaquil, Ecuador

2357 **MATTHEWS, GERALDINE M.** Information center concept in mental retardation. *Association of Hospital and Institution Libraries Quarterly*, 11(3):28-30, 1971.

Although staff or professional library services in MR facilities lag behind other medical library formats, MR professional libraries can make a unique, innovative contribution to library and information services. The MR library design should be interdisciplinary, attuned to the needs of all users, should stress quick and accurate response to all information requests, incorporate audiovisual media, be involved in "in-service training," and provide community services and public relations functions. The MR library must view itself in a broader context than that traditionally assigned to the library and seek to be a library-information service which organizes all information in any medium into the format most accessible to all of its users. (3 refs.) - *S. Glinsky.*

No address

2358 Overview. *Association of Hospital and Institution Libraries Quarterly*, 11(3):27-28, 1971.

Increasingly librarians are realizing the library needs of MRs and are actively promoting programs of library service to MRs. Only recently have librarians recognized their role in the lives of MRs. Prior neglect in this field was due to a variety of reasons, including the inflexible library image (which stressed only reading), a lack of money, and a lack of awareness of MR library needs. The last decade has brought the library into the field of MR both as a repository of newly developed information for the researcher and MR worker and also as a service agency for MR needs. Community library services for MRs have developed according to many formats, and a recent survey discloses that 110 state institutions out of 151 in the United States provide library service for staff and residents. In several states the public library system is responsible for providing materials and services to MR institutions. In the final analysis it devolves upon the practicing librarian to originate a pragmatic response to the informational needs of MRs. (1 ref.) - *S. Glinsky.*

2359 **KLUMB, MRS. DAVID; NAVIS, MRS. RONALD; & BUSSE, MRS. WALTER.** A public library works with the retarded. *Association of Hospital and Institution Libraries Quarterly*, 11(3):32-34, 1971.

A public library staff succeeded in presenting weekly story hours to child and adult MRs. Three Mead Public Library children's librarians organized a weekly story hour for a class of 8 MRs, aged 7 to 9 years, whose mentality was pre-school level. The object of the story hour was not only entertainment but also growth in understanding of the library's uses by the MRs. The program was based on various books and stories and included the use of a flannelgraph, films, filmstrips, puppets, life-sized puppets, records, and live animals. The 31 hour program brought the MRs out of their condition of disinterest and resulted in animated responsiveness, interest, and enthusiasm. The program for the 14 adult TMRs sought to add dimension to their lives and to develop interests and an awareness of their surroundings. This was accomplished by cooperative projects, such as planting seeds, as well as by films, reports on bird-watching, music in films and records, and field trips. Enlistment of community help in additional projects is planned. (1 ref.) - *S. Glinsky.*

No address

2360 **BIALAC, VERDA.** The public library and the retarded patron. *Association of Hospital and Institution Libraries Quarterly*, 11(3):31-32, 1971.

Public libraries have a growing responsibility to help the larger numbers of MRs who are returning to their own families or to community housing. Guidelines for working with adult EMRs include library provision of materials appropriate to MRs, such as books with low-level vocabularies and high-level interest, personal welcoming and introduction of the MR patron to the library, direction of the MR to the library sections containing materials designed for MRs, and explanation of checking out procedures. The MR can and will be a responsible and satisfied patron, if an atmosphere of friendliness and personal encouragement is maintained. (1 ref.) - *S. Glinsky.*

No address

PROGRAMMATIC ASPECTS – Residential

2361 **PEDERSEN, JENS MALLING.** The physical environment of the mentally handicapped. I. Progress in building for the mentally handicapped. *Journal of Mental Subnormality*, 16(2):121-125, 1970.

Architectural planning for the MR should be based on the concept that they have the same needs as other human beings plus a few special needs. The architect must identify the special requirements of the MR by studying the medical-therapeutic-educational work and development and by obtaining feedback on the effectiveness of existing facilities. In a facility for the MR in Jutland, Denmark, each resident has a private room in a series of 10 houses designed

for 24 patients each. The change in environment seemed to make an enormous positive difference in the patterns of behavior of SMRs and PMRs within weeks after their transfer into these houses. The present trend is toward reduction in the size of hospitals for the MR. Future architectural needs will include outpatient facilities, apartments, and hostels. (No refs.) - *J. K. Wyatt.*

No address

2362 **HEATON-WARD, W. ALAN.** The role of the hospital in the mental subnormality service. *Journal of Mental Subnormality*, 16(31):57-61, 1970.

Hospitals for the MR should be a fully integrated part of the Community Service. They should provide residential care for the MR when this type of placement seems the most appropriate. Their services should include assessment, long-term and short-term care, care by an interdisciplinary team, and a basic-care training program for nurses and all those engaged in the care of the MR. Full assessment should be required prior to acceptance for long-term care. The size of new hospitals should be reduced, and older, larger hospitals should be adapted to permit family-type patient grouping. The psychiatric staff should have a primary interest in the field of MR. Hospital isolation and the barriers between the hospital and the community can be reduced by the increased use of volunteers. (3 refs.) - *J. K. Wyatt.*

Stoke Park Hospital Group
Stapleton, Bristol, England

2363 **NIRJE, BENGT.** Symposium on "Normalization." The normalization principle — implications and comments. *Journal of Mental Subnormality*, 16(31):62-70, 1970.

When programming for MRs is based on the normalization principle, developed through work with MR primarily in Scandinavian countries, experiences in the normal rhythm of the day, the week, and the year as well as opportunities to participate in the normal developmental experiences of the life cycle are provided with due consideration to specific individual handicaps. Normal personal considerations are encouraged, and opportunities for bisexual contacts are provided. Normal economic standards are applied so that MRs can learn to cope with economic problems, obtain realistic social training, and gain greater independence. Living facilities are modeled on those of normal society with respect to size, location, integration, and decoration. They provide special features to facilitate the mobility of the physically handicapped. The application of the normalization principle can help many MRs to achieve complete independence and social integration, while others will gain relative independence, depending on their individual needs. In addition to its effects on the lives of the MR, the normalization principle profoundly affects the attitudes of the public, the staff members who work with MRs, and the parents. (8 refs.) - *J. K. Wyatt.*

No address

2364 **GUNZBURG, H. C.** Symposium on "Normalization." II. The hospital as a normalizing training environment. *Journal of Mental Subnormality*, 16(31):71-83, 1970.

When hospitals for the MR are reshaped and reorganized to function as normalized training environments, institutional life is carefully manipulated to offer full preparation for normal life for those who can achieve it, while providing a protective corner within the community. Many varied normal learning opportunities and a climate, environment, and rhythm of living which are as near as possible to normal life are provided. Programming seeks to counteract faulty and inadequate past experiences and provides preparation for comparatively independent life in an open community. Normalized institutions require multidisciplinary leadership, a planned variety of different professional skills, and an integrated training program focused on normal life practices. Work training should be designed to approximate normal work conditions, but should not replace normal community work situations. Levels of training suitable for different levels of capability should be clearly differentiated and should include sheltered shop occupation, training, and work in the community. Social education should be provided in a new type of residential domestic setting, the Living Unit, which provides a "normal approach" to

everyday living problems and direct preparation for life in the community. (15 refs.) - *J. K. Wyatt.*

Hospitals for the Subnormal
Birmingham, England

2365 **ZARFAS, D. E.** Symposium on "Normalization." III. Moving toward the normalcy principle in a large government operated facility for the mentally retarded. *Journal of Mental Subnormality*, 16(31):84-92, 1970.

The Unit System is an application of the normalcy principle in an institution for the MR in Ontario, Canada, which is based on the provision of individualized treatment and training with dignity for living. The goal of the program is to move the institution away from a medical-nursing orientation to a model based on learning-living principles. Residents with similar needs who can benefit from similar programs are grouped together and receive the services of hospital, educational, activity, and/or adult training and rehabilitation units. Coordinators have been added to the Central MR Branch Services Staff to support the director in the areas of community resources, training, and staff development. New staff training programs include a 6-week retraining course for nurses' aides and attendants, and a 2-year, 3-phase program for all newly recruited staff members. Individualization has been easiest in the smaller places where existing staff-resident ratios are higher. Since no standards for staff to resident ratios existed, those of the SAMD were adopted with some modifications. After 2 years of operation, normalization has been approached rather than achieved, and the system is in a constant state of change. (No refs.) - *J. K. Wyatt.*

Department of Health
Ontario, Canada

2366 **HAMMER, N.; TOMKIEWICZ, S.; & DUCHE, D. J.** L'hospitalisation a Paris des enfants arrieres profonds (Hospitalization in Paris of profoundly retarded children). *Bulletin de l'I. N. S. E. R. M.*, 23(6):1355-1450, 1968.

A retrospective study of 1,513 PMR children hospitalized in Paris over a period of 6 years yielded specific social and medical characteristics pertaining to this group of patients. Particular attention was paid to aspects of hospitalization and to the future of these children upon discharge from the hospital. (27 refs.) - *B. J. Grylack.*

I.N.S.E.R.M. Groupe de Recherches
d'Hygiene mentale de l'Enfance et de
l'Adolescence inadaptees
92 - Montrouge, France

2367 **POSTERNAK, YVONNE.** La participation des parents et des volontaires dans nos institutions (The participation of parents and volunteers in our institutions). *Nos Enfants Inadaptes*, 34(2):22-24, 1970.

Special schools, day care centers, workshops, and summer camps for MR children have been established in many countries only within the past decade, in many instances under pressure from parent associations. Residential institutions for MR children often display paternalistic attitudes towards families and may ignore or misjudge the duties and rights of parents. These institutions should adopt a certain form of social integration and should not isolate themselves from the life of the local community. The importance of volunteer organizations is developing in the majority of countries today. Although the utilization of volunteers is not a means of alleviating the shortage of trained professionals, it is a significant way of introducing new ideas and stimuli into specialized institutions. (No refs.) - *B. J. Grylack.*

No address

2368 **POMEROY, DIANA.** Rethinking the bedroom image. *Special Education*, 59(4):24-26, 1970.

A general survey of the living environments of children in 7 schools for the physically handicapped, 4 schools for the deaf and partially hearing, and 1 school each for children with spina bifida, epilepsy, blindness, MR, and emotional maladjustment investigated bedroom areas, staff/child ratios, patterns of care, types of handicaps, number in bedroom group, and time

spent in bedroom area. Conflicting opinions were found regarding heterogeneous ability grouping and mixing children of different ages in a bedroom group. Nealry 25% of the rooms were for 4 children, although number of children in a room ranged from 1 to 26. In 75% of the schools, a limited number of single rooms were available to isolate children with social and behavior problems. Social interaction was greater among smaller bedroom groupings. An in depth study of 6 SMR girls with cerebral palsy (CA range 13 to 16 yrs) revealed that the factors of keeping possessions near their beds, colored bed covers, and low partitions for privacy had important positive effects on social behavior. (No refs.) - *J. K. Wyatt.*

Bristol University
Bristol, England

2369 **MANEY, ANN C.; PACE, ROBERT; & MORRISON, DONALD F.** A note on Shellhaas and Nihira's "Factor analysis of reasons retardates are referred to an institution." *American Journal of Mental Deficiency*, 75(3):316-318, 1970.

Problem areas in the methodology utilized by Shellhaas and Nihira in their "Factor Analysis of Reasons Retardates Are Referred to an Institution" are discussed with a view toward establishing a generalizable body of knowledge. The issues include the choice between a clinical and a statistical focus in the development of data for program planning; the use of principal component analysis as a means of parsimoniously describing population variance in a set of items vs. the use of factor analysis procedures as a means of arriving at conceptual distinctions within a set of items; the use of systematic rather than impressionistic procedures in the comparison of factor structures; and the selection vs. the exclusion of disadvantaged, minority group background as a population parameter in studies of institutional administrations. (5 refs.) - *S. Glinsky.*

National Institute of Mental Health
Adelphi, Maryland 20783

2370 **MEILE, RICHARD L.; & BURK, H. WARREN.** Group relationships among institutional retardates. *American Journal of Mental Deficiency*, 75(3):268-275, 1970.

Peer groups among institutionalized MRs function in a manner similar to those of other institution populations. A sample of institutionalized MRs was studied to ascertain the implications of group structures for adjustment and conformity to institution rules. The hypotheses tested were that group members and isolates would not differ significantly in adjustment to the institution, that group norms would accord with the formal requirements of the "host" organization, and that membership in a peer group of MRs would be a function of sharing common, personal, and social characteristics. The Ss were 85 institutionalized MRs with an ability to verbalize, a CA range of 11 to 30 years, and an equal representation of males and females. Data were gathered by means of personal interviews with both structured and open-ended questions. Results indicated that peer groups functioned to socialize and support members, with members more likely to deviate from formal rules than the isolates. Membership in a peer group was facilitated by similarity to others in personal and social characteristics. Also there was some difference between the sexes in the salience of social-emotional and task functions of their peer group. The findings indicate the need for more research on the nature, function, and implications of peer group relationships among institutionalized MRs. (29 refs.) - *S. Glinsky.*

University of Nebraska
Lincoln, Nebraska 68506

2371 The chaplaincy in institutions for the retarded. *The Scroll*, 9(2):1-6, 1969.

A Joint Accreditation Research Committee of the Religion Subdivision, AAMD, has submitted a planning document outlining the role of religious chaplains in MR institutions. Professionally trained chaplains are regarded as essential to the staff of MR institutions for the religious welfare of MRs and are responsible for the total religious program for the residents. Chaplains must meet minimal training standards as clergymen and in clinical pastoral work. The state has certain obligations, including provision of a place of worship, office space, and secretarial assistance, payment of the chaplain's salary, and securing commensurate professional benefits for the chaplain. Finally, a certification committee is to be set up to administer certification of clergymen as clinical pastors. (No refs.) - *S. Glinsky.*

2372 Evaluation of population characteristics of the Washington State schools for the retarded. *Research Report*, 3(15), 51p., March 1971.

Data presented include an overall view of the population by states in 5 schools for the MR for the fiscal years 1961 to 1970, and a history of the movements over a 10-year period of persons admitted during fiscal year 1961. "On-the-books" population increased by 686 persons during the 9-year period. However, there was an actual decrease of 48 persons in the resident population and an increase of 738 individuals on placement. There was a 292% increase in the number of persons on placement during this time reflecting a 317% increase for females and a 280% increase for males. Placements were generally made for individuals in higher age brackets. Of 377 individuals admitted during fiscal year 1961, 295 were "on-the-books" in 1970 (255 in resident status, 40 in placement status). There was a general annual increase in the percentage of this group on placement status, although the group did not attain the percentage of persons on placement status that was attained for the overall population. (No refs.) - *J. K. Wyatt.*

2373 **ABBAS, K. A.; DONOGHUE, ELAINE C.; & GAL, E.** A computer evaluation of the demand for short-term or long-term care for mentally subnormal children. *Journal of Mental Deficiency*, 14(4):312-318, 1970.

Assessment of the determinants of long-term care and short-term care in 285 MR children who were admitted to a hospital over a 3-year period disclosed that maternal ill health was the only determinant of short-term care in preference to long-term care. Factors significantly related to long-term care which appeared partially modifiable were child management difficulties, total dependency in walking or feeding, open parental rejection, and suspected abnormality at birth. Long-term care was also significantly related to illegitimacy, gross physical anomalies, and inadequate previous placement outside the home, which are factors less likely to be modified. Length of care was not significantly affected by sex, age of admission, mental level, language, emotionally disturbed behavior, medical advice to hospitalize a child, general physical condition on examination, epilepsy, cerebral palsy, Down's syndrome, parent age, social class, housing, number of siblings, or total dependency in dressing or toileting. (13 refs.) - *J. K. Wyatt.*

Queen Mary's Hospital for Children
Carshalton, Surrey, England

2374 **FORSSMAN, H.; & AKESSON, H. O.** Mortality of the mentally deficient: A study of 12,903 institutionalized subjects. *Journal of Mental Deficiency Research*, 14(4):276-294, 1970.

Investigation of mortality data on 12,903 special class or institutionalized MRs over a 23-year period (1955-1968) revealed an overall mortality rate 3.2 times greater than that of the normal population. Reduction of life expectancy was 5% for the whole series, 2% for the mildly retarded, 7% for SMRs, 6% for patients with Down's syndrome, 14% for epileptics, and 6% for hard-to-manage MRs. Relative mortality rates were higher for females than for males, especially among the younger patients. Relative mortality rates for mildly and severely MR individuals were 1.7 and 4.1 times the normal rate, respectively. Excess mortality rates were 9.4 times the normal for Down's syndrome, 7.9 times the normal for epilepsy, and 2.8 times the normal for hard-to-manage MRs. When compared to earlier life expectancy studies, these figures represent a small decrease in average relative survival times. (8 refs.) - *J. K. Wyatt.*

University of Goteborg
S-42203 Hisings Backa, Sweden

2375 **CHANCE, ARTHUR.** Hostels within the community. *Teaching and Training*, 8(4):102-108, 1970.

Hostels for MRs represent an important advance in community care as opposed to hospital care and must be increasingly accepted and supported by the entire community. Institutional care provides only for basic existence requirements, such as food, clothing and shelter, while hostels administer to individual human, emotional needs. The function of hostel wardens is to meet the needs of hostel residents while also promoting professionalism among the hostel staff and improving the status of staff. The aims of hostels

of all types are to furnish as nearly as possible a normal family life for MRs and to rehabilitate the residents for eventual acceptance by the community. Many problems are still attendant upon the development of hostels, including cooperation between community and authorities, as well as between medical and welfare departments. If this instrument is to develop true community care, full cooperation and understanding are required from both officials and the community at large. (No refs.) - *S. Glinsky.*

No address

2376 KRUGMAN, SAUL. Experiments at the Willowbrook State School. *Lancet,* 1(7706):966-967, 1971. (Letter)

Hepatitis experimentation at the Willowbrook State School for the MR can be defended on the basis that viral hepatitis is so prevalent that newly admitted susceptible children become infected within 6 to 12 months of entry and that recent studies on active and passive immunization for the prevention of viral hepatitis, type B, have demonstrated clearly a therapeutic effect for the children involved. The proposal to expose a small number of newly admitted children to the strains of hepatitis virus endemic to Willowbrook was justified by the inevitability of exposure of these children, the admission of the children to a well-equipped unit where they were isolated from other infectious diseases, the likelihood that they would have a subclinical infection as a result of the experiment which would be followed by immunity, and the receipt of informal consent from parents of all participating children. (2 refs.) - *B. J. Grylack.*

New York University School of Medicine
New York, New York 10016

2377 SPENCER, D. A. Education for autistic children. *Lancet,* 2(7686):1309, 1970. (Letter)

The fact that 1 out of every 7 autistic children is placed in mental-subnormality hospitals due to a lack of other residential accommodation underlines the dilemma facing these hospitals with regard to autistic children. Even though doctors at these hospitals consider such accommodation unsuitable, other segments of the community argue that autistic children who have made little progress at special schools are candidates for admission to these hospitals since they are functioning at a subnormal level. While hospitals are criticized for excluding these children, they are also condemned for admitting them. (No refs.) - *B. J. Grylack.*

Meanwood Park Hospital
Leeds 6, England

2378 DONOGHUE, E. C.; ABBAS, K. A.; & GAL, E. "Put away"—for how long? *Nature,* 227(5261):972, 1970. (Letter)

The present tendency in England is for subnormality patients to remain hospitalized for short periods of time; most (80%) remain at home. In one hospital, of 164 short-term children admitted, 9% stayed longer than 8 weeks. There is virtually no current option at this institution for long-term stay. There were no significant psychological differences between long-term and short-term patients. (10 refs.) - *E. Kravitz.*

Queen Mary's Hospital for Children
Carshalton, Surrey, England

2379 SPENCER, D. A. Strategy for hospitals for the mentally handicapped. *Lancet,* 1(7701):697-698, 1971. (Letter)

Hospitals for the MR are not inherently bad and can be made both economical and effective. Patient classification, outpatient services, community-related programs, controlled admissions, a pleasanter environment, specialized forms of therapy, and better public relations can all contribute to the development of facilities which will be truly beneficial to patients. (No refs.) - *E. L. Rowan.*

Meanwood Park Hospital
Leeds 6, England

2380 Kinsmen Elmwood Lodge, Saskatoon. *Deficience Mentale/Mental Retardation,* 21(2):16-19, 1971.

The Elmwood concept primarily tries to provide an answer to the question 'where shall my retarded child live.' In 1966 the Saskatoon Branch of the Canadian Association for the Mentally Retarded attempted to find its own answer through the creation of a residence in the city. Through the involvement of citizens and the financial assistance and cooperation of the central government and City Council, in 1969 the opening of a residence which could accommodate 50 MR children in a home-like atmosphere was accomplished. (No refs.) - *P. Custers.*

2381 The message of Farleigh. *Lancet*, 1(7703):790-791, 1971. (Editorial)

A committee of inquiry appointed to investigate allegations that mentally handicapped patients were being mistreated by male nurses at Farleigh Hospital near the village of Flax Bourton in England issued its report in the spring of 1971. The committee found that protracted administrative disputes, inadequate hierarchical structures among the staff, and a total lack of psychiatric leadership, combined with the severe demands of nursing care and deplorable physical conditions on the male ward, led to the abuses. The committee recommended that: staff structures should be revised to minimize the isolation of chief nursing officers; an organizer of volunteer services should be appointed; a code of conduct for handling violent patients should be formulated; a practicing psychiatrist should be appointed to every regional hospital board; there should be more adequate financing for such institutions; and a Health Commissioner with investigative powers over the National Health Service should be appointed. Nurses who undertake care of the grossly handicapped are entering an arduous service and deserve the full support of doctors and society in coping with its difficulties. Free and full discussions involving all those responsible for the care of the MR are needed. (No refs.) - *N. Jarvis.*

2382 **BUSHNELL, ROGER K.** Bevan Lodge, Courtenay, B.C. *Deficience Mentale/Mental Retardation*, 21(2):14-16, 1971.

Bevan Lodge is a residential facility for 74 adult retarded persons, with a stimulating environment inherited from its previous function as a summer resort. Since its establishment in 1968, Bevan Lodge has enlisted excellent community response from the surrounding area. The philosophy of the residence is to promote the happiness of each resident by showing respect for the person as an individual, believing in his ability to decide his own lot, and helping the adult retarded to discover his untapped capacities. The overall aim of the Lodge community is to create a family situation with shared responsibilities. (No refs.) - *P. Custers.*

Bevan Lodge Association
Courtenay, British Columbia, Canada

2383 **POSTERNAK, YVONNE.** La participation des parents et des volontaires dans nos institutions (The participation of parents and volunteers in our institutions). *Deficience Mentale/Mental Retardation*, 21(2):44-46, 1971.

Parents of mentally retarded children need to be involved in the administration and program planning of the various institutions which exist to meet their children's needs. In recent years parent associations have become increasingly involved with the organization of different institutions for their children, such as schools, summer camps, workshops, etc. This parental involvement should now be extended to residential institutions so that family members might continue to contribute to planning for their child's welfare. Adequate transportation needs to be provided, and family members must be made to feel that their contributions are valuable. Realizing this possibility demands a substantial change in the current attitudes of MR institutions toward parent participation. Recognition of the potentially valuable roles to be played by parents and other volunteers as important links to the larger community should facilitate this needed change in attitude. (No refs.) - *P. Custers.*

No address

2384 **RICHARDSON, B.** Salmon in a psychiatric hospital group. 4. The Salmon unit in subnormality. *Nursing Mirror*, 131(21-24):23-24, 1970.

A Salmon unit may be helpful in improving the presently inadequate hospital care of mentally

retarded patients. Such a unit must be set up empirically since there are not enough experimental data to assist. The system described segregates male, female, and children. Teaching situations are utilized as they present themselves. An attempt is made to avoid those aspects of institutionalization, e.g., depersonalization and block treatment, which tend to reinforce existing illness. Regular unit meetings encourage pertinent input from various personnel. Unit housekeeping is preferred. (No refs.) - *E. Kravitz.*

Leicester Frith Hospital
Leicester, England

2385 **BARTZ, WAYNE R.; LOY, DONALD L.; & COOK, WILLIAM A.** Mental hospitals and the winds of change. *Mental Hygiene*, 55(2):266-269, 1971.

The state hospital of the future will be limited to the care of special patients who cannot be accommodated in community mental health centers. A philosophical shift from general treatment to total control will be necessary in order to handle alcoholics, drug users, acting out adolescents, MR individuals with behavior problems, chronic patients without families, patients with character disorders, and individuals dangerous to others. (2 refs.) - *E. L. Rowan.*

Dewitt State Hospital
Auburn, California 95603

2386 **GARDNER, JAMES M.** Innovation in the delivery of psychological services in an institution. *American Psychologist*, 26(2):211-214, 1971.

When attendant level personnel were utilized in a more responsible manner, there was an increase in the adaptive level of MR children in a state institution. Attendants were allowed to operate in their own style and were given responsibility for the progress of individual patients. Feedback from supervisors in a group setting was most effective in improving attendant skill and personal growth. The patients' growth was directly related to that of the attendant. The professional staff members were free to function in their areas of special competence. (No refs.) - *E. L. Rowan.*

Orient State Institute
Orient, Ohio 43146

2387 Utopian village for the mental handicapped? *Journal of the Irish Medical Association*, 64(406):121-122, 1971.

Among subjects discussed at the fourth Lota seminar for the mentally handicapped in Cork, Ireland, were the incidence of SMR children; a residential hostel in Southampton that provides facilities for 24 SMR children; obstacles to effective care, such as rigid routine, block treatment, depersonalization, and social distance; the necessity for a reduced staff-patient ratio and the willingness of everyone to perform assigned duties to achieve successful care of the MR. (No refs.) - *J. K. Wyatt.*

2388 **HARDLEY, P.** Short-term care for mentally handicapped children. *Nursing Mirror*, 132(16):34-39, 1971.

Hill House Hospital provides a well-articulated short-term care program for MRs to an age limit of 12 years. Reasons for admission include assessment of the present condition and future needs of the MR child, relief of strain upon the MR's family, and provision of opportunity for the MR's family to go on vacation and receive hospital treatment. The short-term care unit of the hospital has 14 beds for short-term patients and 138 beds for informal patients, as well as experienced nurses and cadet nurses. Admission to the hospital follows upon application for admission to local health authorities and prior examination of the MR by a general practitioner. A parental interview determines the particular needs of the MR and establishes mutual confidence in parents and the MR. Routine procedures consist of a physical and neurological examination with subsequent consultation with a psychiatrist and other supporting specialist staff. The daily schedule centers around both morning and afternoon sessions in the hospital school, interspersed with and followed by play periods. Disadvantages of short-term care, such as deprivation of family care and interruption of progress by the staff in understanding and helping the MR, are far outweighed by advantages, including relief of pressure on parents, prevention or delay of permanent admissions, and advice on home care problems given to parents. (No refs.) - *S. Glinsky.*

Hill House Hospital
Rye, Sussex, England

2389 **LIEBIG, MARGARET.** Direct service to residents in an institution. *Association of Hospital and Institution Libraries Quarterly*, 11(3):34-35, 1971.

The library service program at the Central Wisconsin Colony and Training School has been successful in reaching the SMR and PMR. The program is designed primarily to help all residents develop their fullest potential and is also a materials center for the ward programs, the school, and other departments. In addition to books, the library provides story hours and utilizes filmstrips, records, cassette tapes, puzzles, games, and toys. Story hours succeed best when stories are told which demand motor involvement, such as clapping, singing, or rhythmic movement. The library also circulates a book truck to wards once a week for patients who cannot come to the library. This personal service is well received and has also stimulated ward staff utilization of the library's resources. (No refs.) - *S. Glinsky.*

Central Wisconsin Colony and
Training School
Madison, Wisconsin 53704

2390 Recommended standards for library services submitted by the committee on library services to the accreditation council for facilities for the mentally retarded (unapproved draft 1/7/71). *Association of Hospital and Institution Libraries Quarterly*, 11(3):36-38, 1971.

A series of 12 major guidelines outlines recommended standards for library services for MR facilities. Library services shall be available to the facility to support its total rehabilitation program and provide multimedia information to both staff and residents, regardless of age, IQ, or handicap. Services to residents should include assistance in team evaluation of individual MR's developmental level in communication; provision of appropriate informational, recreational, and educational materials; and development of various levels of programs for residents. Library services to be provided to the staff are to be appropriately detailed in written policies and properly staffed and incorporated into the overall organizational structure of the facility. Services rendered outside the facility must be rendered according to formal agreement regarding lines of communication, areas of responsibility, and kinds of service. Finally, the professional and academic standards of all library personnel are detailed. (No refs.) - *S. Glinsky.*

2391 **FORMAN, MARK.** The alienated resident and the alienating institution: A case for peer group intervention. *Social Work*, 16(2):47-54, 1971.

Peer group intervention between an alienated resident and an alienating institution is at times appropriate. Mutual alienation between the institution and residents impairs the ability of both to recognize or work on common goals. Resident groups provide a meeting ground where the needs of residents and of the institution alike can converge. Although the function of such intervention groups in an adolescent MR institution is discussed, the implications of this study are valid for all MR institutions. (13 refs.) - *S. Glinsky.*

Rutgers—The State University
New Brunswick, New Jersey

2392 **MITRA, SUDHANSU B.** Guidelines for hospitalized retarded-deaf children. *American Annals of the Deaf*, 116(3):385-388, 1971.

A set of guidelines for program planning for institutionalized MR deaf children is proposed. The guidelines include program objectives, criteria for selection and grouping of children for instructional purposes, and basic arts and skills pertaining to hearing impairment and MR. In addition to standards for teacher selection, essential provisions for developing a program are suggested. Program evaluation should emphasize placement and followup. (9 refs.) - *S. Glinsky.*

Coppin State College
Baltimore, Maryland 21216

PROGRAMMATIC ASPECTS – Recreational

2393 **BLID, EVY; & JUNKER, KARIN STENSLAND.** La creation d'une "lekotek" (The creation of a "toy library"). *Nos Enfants Inadaptes,* 34(2):20-21, 1970.

The principle of a "toy library" is to furnish toys and education materials appropriate for mentally handicapped children at each level of development. Parents arrive with their retarded child for sessions of approximately 2 hours in length. Here they receive counseling on activities that will aid the development of their child, and the child benefits from the devotion of this time exclusively to his needs. Special exercises are employed with the purpose of expanding future possibilities for the child. It is hoped that successful experiences with this new program will encourage the manufacture of toys and educational materials which will contribute to the child's psychomotor and intellectual development. (No refs.) - *B. J. Grylack.*

No address

2394 **BYLES, THEA.** Some observations on the teaching of nature study to severely retarded teenagers in a rural area. *Teaching and Training,* 8(4):113-114, 1970.

The study of nature is a valuable tool in the development of SMR teenagers. Nature furnishes a multitude of sensory experiences, provides a basis for growth in language and concept development, gives enjoyment, which further enhances learning motivation, and teaches esthetic values. A most practical adjunct for nature study is a nature table for displaying items found by the SMRs on nature walks. This gives opportunities for labeling and writing, as well as artistic activities. Nature study attunes the SMR to his natural surroundings and opens the way to other interests and eventual integration into the community. (No refs.) - *S. Glinsky.*

Attleborough Training Centre
Norfolk, England

2395 **ROURKE, R. J.** A share of summer fun. *Journal of Health Physical Education Recreation,* 42(5):71-72, 1971.

The Summer Recreational Camping Program of New York State's Division of Mental Retardation provides a 2-week outdoor educational summer camping experience for more than 2,000 carefully screened retarded children and adults that improves relations between state schools and communities, provides employment and opportunities for young career-minded persons, and enhances the retardate's personal development. Presenting a new and stimulating sociorecreational experience tailored to participants' needs, the program provides hikes, swimming, boating, song festivals, ball games, and numerous activities focused on distinct themes. Professional administrators and college student counselors furnish closely supervised learning in daily-living skills, human interaction, and various activity periods grouped under 4 categories: arts and crafts, music and drama, recreation and nature, and waterfront. The consensus is that the investment has been returned manyfold. (No refs.) - *B. Berman.*

Division of Mental Retardation
Albany, New York

2396 **BREWER, FRAN.** Philosophy of recreation for the mentally retarded. *Journal of Health Physical Education Recreation,* 42(5):76, 1971.

Recreation for the retarded is a necessary, universal, and deserving service that provides away-from-home, democratic experiences, induces respect for others, stimulates imagination, creativity and interest in nature, and furnishes fun and a focus for building ideals and educational growth. (No refs.) - *B. Berman.*

Florida State University
Tallahassee, Florida

2397 BAKER, CHARLES. Therapeutic recreation for exceptional children. *Journal of Rehabilitation*, 37(1):32-34, 1971.

It is finally being recognized that recreation can be used effectively as a therapeutic tool in the treatment of children who suffer from disabilities. Recreation constitutes an important means of dealing with the lower attention tolerance level in retarded children and is also a significant medical adjunct in the preservation of physical and mental health. Recreation has boundless potential as a therapeutic tool. Play can preserve the uniqueness of each retarded child and can help to reveal hidden capabilities and to develop them to the fullest possible extent. (7 refs.) - *B. J. Grylack.*

Protestant Home for Children
Buffalo, New York

2398 ANDRES, CATHERINE. The status of municipal recreation for the mentally retarded. *Therapeutic Recreation Journal*, 4(1):inside cover, 30, 1970.

In April, 1967, 166 of the largest cities in the United states were surveyed with regard to municipal recreation programs for MRs. The results obtained from the 121 returned questionnaires indicate a probable increase in the number of these programs, but too many departments are offering programs only for children and are overly dependent upon volunteer agencies to provide recreation for MRs. Although improvement is evident, there is still a lack of programs designed for MRs even in the larger cities. Recreational activities are important to ensure maximum growth and development. Municipal recreation departments have the necessary professional staff and community support to provide these activities, and they must begin to assume the initiative in including both MR children and adults in their programs. (No refs.) - *B. J. Grylack.*

Childrens Orthopedic Hospital
Seattle, Washington

2399 CLOT, M. Une colonie de vacances de l'A.D.A.P.E.I. de l'Isere (A holiday camp run by the A.D.A.P.E.I. of Isere). *Nos Enfants Inadaptes*, 36:12-16, 1970.

From July 31, until August 29, 1969, a holiday camp was organized for 63 mentally retarded children, under the guidance of a team of 24 persons, the majority of whom had had no previous experience in care of the MR. Apart from many daily problems, major attention was given to the organization of special events outside the camp, such as swimming in a local pool, picnics, attendance at a circus, and a special party, all of which required a continual re-organization of group structures in the camp. A note of self criticism concerns the inadequate preparation of the supervisors in pre-knowledge of the individual children. It is suggested that this situation be improved upon in future camp planning; specific recommendations are also made with respect to improved staff organization. (No refs.) - *P. Custers.*

No address

2400 WEST, JENNY. Horse riding for mentally handicapped children. 131(12):13-14, 1970.

The value of carefully supervised horse riding for certain mentally handicapped children is discussed. Patience, understanding, and expert guidance are needed. Horse riding has proven to be of physical benefit and has offered a stimulating and satisfying activity to these children. There are more than 70 groups with room for 2,000 riders in England, 20% of whom are mentally handicapped. There is an active exchange of ideas and information. (No refs.) - *E. Kravitz.*

No address

2401 VANCON, NANCY. Space to grow. *Rehabilitation Around the World*, 21(5,6):10-15, 1970.

An outing by 30 handicapped children in the Washington National Forest of Virginia is described. Most of the children were mentally retarded. Music, artwork, mineralogy, and special trails for the handicapped were utilized to expand the world of these children. Facilities for the handicapped are being built and enlarged at other parks. About 40 older handicapped adult workers are employed by the United States Forest Service. (No refs.) - *E. Kravitz.*

No address

FAMILY

2402 **CLANCY, HELEN.** A group family holiday: An innovation in the therapeutic management of the autistic child. *Slow Learning Child,* 17(3):149-162, 1970.

To help remove feelings of family isolation and helplessness in handling problems associated with autistic children, 3 holiday workshops were organized for 11 families – each with an autistic child. Guided by voluntary workers, the workshops provided a needed holiday for the families, demonstrated how parents can be mutually helpful, and induced more normal behavior in the autistic children (one previously non-verbal child began to use speech). The young professional volunteers returned to their work with increased understanding of autism, and parents found themselves later applying the insights they had received. The workshops seem valuable for other handicapped children, such as MRs and the deaf. (5 refs.) - *B. Berman.*

University of Queensland
Brisbane, Australia

2403 **WAECHTER, EUGENIA H.** The birth of an exceptional child. *Mental Health Digest,* 2(12):9-12, 1970.

When a newborn baby deviates drastically from the anticipated child, the loss of the idealized child and the demand to cope with the abnormal child can be overwhelming. The manner in which the family deals with the situation is influenced by the way in which professional personnel work with the parents immediately following the birth of the exceptional child. Nurses may play a central role in helping both baby and parents. A nurse who is aware of her own feelings and values, who recognizes the significance of such an experience, and who can contribute her own strength and empathy has a great opportunity to contribute to the comfort and unity of the family and to help create an optimal developmental environment for the child. (No refs.) - *B. J. Grylack.*

No address

2404 **ROUTH, DONALD K.** MMPI responses of mothers and fathers as a function of mental retardation of the child. *American Journal of Mental Deficiency,* 75(3):376-377, 1970.

Analysis of variance of the Minnesota Multiphasic Personality Inventory (MMPI) scales of parents of MR children attending a child development clinic revealed no significant effects of the MR of the child, of the child's age, or of the interactions of the variables studied. The Ss were 48 parents with MR children. The former were randomly picked out of 700 files over the period of a year and administered the MMPI measures. The study did not fulfill predictions, although parental education was significantly related to several MMPI scales of each parent. This lack of fulfilled predictions can perhaps be linked to the kind and nature of the controls applied. (7 refs.) - *S. Glinsky.*

Bowling Green State University
Bowling Green, Ohio 43402

2405 **KERSHNER, JOHN R.** Intellectual and social development in relation to family functioning: A longitudinal comparison of home vs. institutional effects. *American Journal of Mental Deficiency,* 75(3):276-284, 1970.

Family functioning, social quotient (SQ), and the relationship of SQ to IQ are factors significantly related to a family's decision to admit an MR child to an institution. A sample of children who were to be institutionalized and a comparable sample who were not to be placed were studied

to determine child and family factors in the placement decision and to ascertain the comparative effects of longterm residential versus home care on selected child and family factors. The Ss were 69 MRs, of whom 42 were institutionalized and 27 remained in the community. The groups were of Caucasian origin and matched by CA and MA. The 42 MRs were tested prior to entry into an institution and one year later by means of SQ and IQ measures. Their families were pre- and post-interviewed to assess the adequacy of family functioning. The factors of family functioning, SQ, and IQ/SQ discrepancy were found to be related to family decisions to seek institutional placement. Community families and children showed decreases over the year on all measures taken, whereas institutional families tended to improve in functioning. Significant positive correlation between IQ decrement and family pretest and posttest losses was found in the community group. In the institution group significant negative correlation appeared between low initial SQ and family increments in functioning. The results underline the important reciprocal relations existing between the MR and his family. (32 refs.) - *S. Glinsky.*

University of California, Los Angeles
Los Angeles, California 90024

2406 **COX, ALICE M.** Le retard du developpement et de l'organisation des fonctions mentales chez l'etre humain. II.—Les fondements familiaux du retard mental. (Revue choisie de la litterature mondiale) (Retardation of the development and organization of mental functions in the human being. II.—The familial foundations of mental retardation. [Selected review of the world literature]). *Annales Medico-Psychologiques,* 2(5):649-695, 1970.

Even in the PMR child, measurement of intellectual potential cannot be achieved in any significant manner without consideration of environmental stimulation. Parental attitudes upon the discovery of MR in their child range from resignation to rejection to overcompensation. Parents who express these reactions in the presence of MR in the family are actually manifesting problems which existed before the birth of the MR infant. The difficulties which parents encounter in raising a retarded child have an inevitable influence upon their attitudes towards the child himself. Sociopsychological research has shown that the attitudes of mothers of MR children are affected primarily by the intellectual capacity of these children. Parents who guide and stimulate MRs effectively have been found to raise unusually gifted children. The psychologist is an indispensable person in helping parents to treat the handicap realistically and in aiding MR children to deal with behavioral and emotional difficulties, some of which may result from the demands of society. (114 refs.) - *B. J. Grylack.*

Indiana State University
Terre Haute, Indiana

2407 **SIEGEL, BESS; SHERIDAN, KATHLEEN; & SHERIDAN, EDWARD P.** Group psychotherapy: Its effects on mothers who rate social performance of retardates. *American Journal of Psychiatry,* 127(9):1215-1217, 1971.

In a community-based day center for the education and training of moderate and severe MRs, 19 mothers of retardates participated in group psychotherapy and 19 other mothers of retardates acted as controls in an evaluation of the effect of therapy upon the social performance rating given by mothers of MRs. After 12 sessions, mothers rated their children according to variables which included listening in a group, truthfulness, obedience, stability, security, conversation, appearance, response to authority, response to criticism, and response to affection. The results show a general trend for the experimental group to rate their children higher on security and appearance after therapy than before it, in contrast to the control group. In view of the significant changes in these 2 ratings, it seems that mothers in group therapy experience greater acceptance of their children. (10 refs.) - *B. J. Grylack.*

Oakland Counseling Service
Pontiac, Michigan

2408 **WILSON, ARTHUR L.** Group therapy for parents of handicapped children. *Rehabilitation Literature,* 32(11):332-335, 1971.

In an attempt to treat the needs of parents of handicapped children, a series of 8 group therapy

sessions was conducted for them under the sponsorship of the local Easter Seal Society and a Title VI Regional Special Education Center. The parents who attended had children with cerebral palsy, blindness, deafness, spina bifida, MR, or muscular dystrophy. Sessions were opened by general remarks from the group leader, who was the parent of a handicapped child and a trained psychologist. This statement of his own experiences soon impelled each parent to recall similar traumas, disappointments, and frustrations in dealing with his child's disability. During the last 3 sessions, specialists in the areas of transportation, MR, and social agency services were invited to attend and speak. Therapy group sessions were conducted simultaneously with a recreation program for the handicapped children of these parents. On the basis of this experience, it appears that parents of handicapped youngsters could form and lead similar therapy groups after appropriate instruction and guidance. (8 refs.) - *B. J. Grylack.*

Fairborn Board of Education
Fairborn, Ohio

2409 **SHARLIN, SHLOMO A., & *POLANSKY, NORMAN A.** The process of infantilization. *American Journal of Orthopsychiatry*, 42(1):92-102, 1971.

Some dynamics of infantilization were investigated within a group of borderline and mildly retarded children in rural Appalachia by interviewing 52 mothers about their child-rearing practices. Infantilization is defined as encouraging another to become or to remain less competent and self-sufficient than he might otherwise be. The operational definition of "having been infantilized" was an IQ loss greater than the average in the sample. Two concurrent measures significantly related to IQ decrement were poorer large muscle coordination and visual clinging to the mother in a task situation. The drop in IQ could be predicted from a composite scale of child-rearing practices. The infantilized children had mothers who more often transmitted 3 messages: "You are part of mother," "You are fragile," and "You are special." The mother unable to separate herself from her retarded child may well exacerbate the existing handicap. Those children did better whose mothers were even-handed and realistic in assessing their limitations, who shared the child with others, and who could be firm and even use spanking when indicated. Two operating mechanisms in the dynamics of overconcern are (1) maternal overprotection as a reaction formation against rejection of the child, and (2) separation anxiety in which the mother clings to her child in a direct expression of her own horror at loneliness. The research further suggests an intergenerational "cycle of infantilization" among mothers. (15 refs.) - *J. C. Moody.*

*University of Georgia
Athens, Georgia 30601

2410 **JOHNSON, MARGARET ANNE.** How mental retardation affects family relationships. *Deficience Mentale/Mental Retardation*, 21(3):38-39, 1971.

Within the family of a retarded child, the manner in which each member deals with the handicap will affect the other individuals in the unit. Arguments concerning the advantages and disadvantages of institutionalizing the child as well as the physical and mental fatigue caused by caring for the child may provoke marital discord. Brothers and sisters are often resentful of the added attention devoted to the handicapped child. A good adjustment by the parents will engender healthy attitudes on the part of siblings. In the adjustment process, development of the ability to face reality is followed by acceptance of the child's mental handicap, constructive steps towards helping him, and efforts towards solving the problems of MR in the community. (No refs.) - *B. J. Grylack.*

No address

2411 **COLLYER, KATHY; & INGRAM, IRENE.** Letters from Nancy's two mothers. *Deficience Mentale/Mental Retardation*, 21(3):27-29, 1971.

A 9-year-old mongoloid female child lived in her original home until a place became available for her in a special school for MRs in another town. This change required that she live in a strange home for most of the school year, spending only occasional weekends with her real family. Initial reluctance by the girl and her family was overcome without significant resentment or depression through the ready adjustment of the foster

family. Through special education, the child has been able to gear her hyperactivity to more constructive endeavors, and she has improved her behavior and increased her trustworthiness. She has retained the affection of her own family in addition to gaining the love of her adopted family. (No refs.) - *B. J. Grylack.*

No address

2412 **KATZ, ELIAS.** *The Retarded Adult at Home: A Guide for Parents.* Seattle, Washington, Special Child Publications, Inc., 1970, 185 p. $3.95.

This basic guide for parents of the mentally retarded responds to frequently asked questions about the problems faced by the adult MR who attempts to lead as independent and normal a life as possible. Sections are devoted to job hunting and preparation, marriage, recreational activities, and problems encountered in community attitudes toward the MR. Suggestions are also made as to practical provisions parents might make to assure continued care for their children after their own death. Appendixes list sources of further information on the availability of financial support and specific community programs for the retarded, and recommend various books, films, organizations and government agencies concerned primarily with problems of the MR. Additional consultation with trained counselors is seen as essential. - *N. Mize.*

2413 **GROSSMAN, FRANCES K.** *Brothers and Sisters of Retarded Children: An Exploratory Study.* Syracuse, New York, Syracuse University Press, 1972, 249 p. $9.50. (Monograph)

A psychological and sociological study of college students with retarded siblings attempts to discover how normal siblings cope with a retarded brother or sister in the family. The sample grouping of college students is divided into "Private University Families" (primarily upper class) and "Community University Families" (chiefly middle class) to account for the influence of socioeconomic standing on family life styles. Findings indicate that sibling reactions to a handicapped brother or sister vary enormously with differences in cultural, socioeconomic, and psychological factors, and do not seem to be significantly determined by the nature or degree of the handicap itself. Most important is the family's general ability to cope with stress, but such variables as sex of MR and normal siblings, age of both, and size of family contribute significantly to the prospects for a positive adaptation. The effect on normal siblings of institutionalization, as opposed to home care, of the MR is also examined with reference to specific clinical material. - *N. Mize.*

2414 **NOLAND, ROBERT L., ed.** *Counseling Parents of the Mentally Retarded: A Sourcebook.* Springfield, Illinois, Charles C. Thomas, 1970, 404 p. $9.75.

A series of articles, previously published over a 20-year period in a variety of journals, have been collected into one work intended as a sourcebook for use in counseling parents of the MR. The book is divided into 6 sections: part 1 provides an overall introduction to the subject, concentrating specifically on the personal feelings and attitudes of the affected parents, as they themselves have expressed them; part 2 discusses alternative approaches to the initial informing interview, and contains several articles on this first session, on counseling during the usual crisis reaction, and on approaches to short-term psychological counseling; part 3 is devoted to group counseling and parent therapy; part 4 discusses the optimal role played by caseworkers in their contacts with families of the MR, and investigates some of the more common personal problems involved in the difficult parental decision to institutionalize their retarded child; parts 5 and 6 are on pastoral counseling and genetic counseling, respectively, with some specific medical information on the hereditary nature of certain types of mental deficiency included in the last section. Appendixes list major parent associations, clinical programs for MR children, and suitable audiovisual material on the subject of MR generally. - *N. Mize.*

2415 **JEFFREE, D. M.; & CASHDAN, ASHER.** The home background of the severely subnormal child: A second study. *British Journal of Medical Psychology*, 44:27-33, 1971.

A follow-up study on the home background of the severely subnormal child, matching 28 children with normal children for mental age, sex, and socioeconomic background (Ss lived at home, but were enrolled in day training centers) revealed, following parental-interview assessment, significant differences overall and in affection, but not in stimulation. Clear differences overall showed on 4 objective tests of experiences as well as on 3 taken individually. (20 refs.) - *B. Berman.*

Manchester University
Manchester, England

2416 **WADSWORTH, H. G.; & WADSWORTH, JOANNA B.** A problem of involvement with parents of mildly retarded children. *Family Coordinator*, 20(2):141-147, 1971.

Questionnaires sent to parents of mildly retarded children in special education classes (as part of a project to involve parents in their children's education) disclosed significant differences between respondents and nonrespondents in socioeconomic status and children's IQ level. Responses also revealed intense feelings toward the problem being investigated and toward classificatory labeling of children, and consequently reinforced already planned school changes. There is a strong need for a parent-education program to induce increased involvement and to dispel many persisting misconceptions about retardation. (No refs.) - *B. Berman.*

Ridge School
Elk Grove Village, Illinois

2417 **MAMULA, RICHARD A.** The use of developmental plans for mentally retarded children in foster family care. *Children*, 18(2):65-68, 1971.

The use of personal developmental plans for EMRs in foster families seems to reduce domination tendencies in the foster mother and to involve the latter more intensively in the MR's development. The Ss were 27 EMRs ranging in age from 6 months to 20 years who were placed in family care. Developmental plans were constructed for 15 children, while 12 children had no developmental plans. Twelve foster mothers or family caretakers were involved, of whom 6 had developmental plans and 6 had none. Each plan was designed to provide concrete immediate and long-range objectives in simple language, guidelines for expected problems, and measures for evaluating the EMRs' progress. It was constructed by the hospital ward staff with the aid of the MR's family. MR evaluation was made every month for 6 months as well as evaluation of the family caretaker's self-image in terms of love and dominance. The results indicated appreciable progress in the cases with plans, with one exception, and an increase in love over dominance in the developmental plan group mothers. Dominance increased over love in the control mothers. The experiment showed that the use of a developmental plan focused the attention and efforts of all involved upon the real, specific needs of the child and accelerated the MR's progress. (3 refs.) - *S. Glinsky.*

California State Department of Social Welfare
Fresno, California

2418 **DEATON, JOHN G.** Managing the retarded child—and his parents. *Hospital Physician*, 7(5):70-75, 1971.

The older Pavlovian advice to institutionalize any MR child as soon as possible is no longer in favor in modern medical thinking. When retarded children are reared at home, where the needed love and attention are more readily available, they achieve higher intelligence and social levels than do their institutionalized counterparts. Medical treatment of MRs differs from treatment of normals only in that it requires more patience. Greater emphasis should be given to proper counseling of the family. Outdated terms such as idiot, imbecile, and moron should be avoided and replaced with the more specific and less offensive categories of mild, moderate, severe, and profound MR. An important objective of genetic counseling is to alleviate parental guilt feelings arising from their having produced a retarded child. (No refs.) - *S. Glinsky.*

Mexia State School
Mexia, Texas

2419 **O'CONNOR, WILLIAM A.; & STACHOWIAK, JAMES.** Patterns of interaction in

families with low adjusted, high adjusted, and mentally retarded members. *Family Process*, 10(2):229-241, 1971.

Families with low adjusted, high adjusted, and MR members showed varying patterns of interaction. Family interactions were compared from families with a low adjusted child prior to formal clinic contact (LA), with a high adjusted child (HA), and with a retarded child (RM). Nine measures were employed, including adaptation, stability, productivity, specificity, overt power, conflict, cohesion, and emotionality. The result suggested a failure of cohesion in the LA group, but many clinic family stereotypes were supported. HA families showed both high cohesion and high conflict. RM families exhibited cohesion, but also many auxiliary control features, such as giving the MR child the youngest sibling role. (10 refs.) - *S. Glinsky*.

Osawatomie State Hospital
Osawatomie, Kansas 66064

PERSONNEL

2420 **DARGASSIES, SAINT-ANNE.** Les problemes sociaux aigus poses par le nouveau-ne pathologique (Acute social problems posed by the sick neonate). *Nos Enfants Inadaptes,* 34(2):11-14, 1970.

Immediate social problems posed by sick neonates are often difficult to resolve, since the medical specialist, occupied with intensive care problems during the neonatal period, is restricted by the absence of social structures which should assist the families of seriously handicapped infants. It is left to the physician to limit the psychic trauma which may arise from the separation of the infant from his family, and from his institutionalization. For the physician, the infant in his care becomes an individual case, and he strives to comprehend how the family of the neonate will be able to bear the immense burden of a retarded child in the future. Although the retention of the infant in the hospital for further study alleviates the weight of uncertainty for the family, it may deprive another neonate who has urgent need of intensive care. Moreover, to succeed in keeping a retarded neonate alive is not the same as to help the family to raise and educate the child. Unfortunately, specialized centers do not always respond to the immediate needs of the family. (No refs.) - *B. J. Grylack.*

Centre de Recherches Biologiques Neonatales de
l'Association Claude-Bernard
Paris 14e, France

2421 **WYNE, MARVIN D.; & SKJEI, PRISCILLA.** The counselor and exceptional pupils: A critical review. *Personnel and Guidance Journal,* 48(10):828-835, 1970.

The counseling profession generally has ignored retarded and disturbed children. Counselors need special knowledge and personal contacts to permit understanding and acceptance of the exceptional child, and to allow them to become resource persons for the classroom teacher. They need training in role playing, group techniques, the use of attitude scales, sociograms, and self-concept measures. More emphasis is needed on guidance and counseling for the EMRs in public schools, since research has shown that students with IQs of 90 or below constitute a different school population in terms of certain non-intellective measures. Structured, directive counseling contacts with 9 low-achieving grade-school children (IQs 80-95) for 10-minute sessions twice a week over 1 academic year produced improved academic achievement and school attendance. Other studies have shown group counseling to contribute to better deportment in MR boys. Crucial in counseling are the overall changes induced in the client. The counselor has a service to offer the retarded child and more information is needed on ways to improve this relationship. (19 refs.) - *B. Berman.*

University of North Carolina
Chapel Hill, North Carolina

2422 Occupational therapy assistant: Creative skills rehabilitate the disabled. *Occupational Outlook Quarterly,* 14(4):12-13, 1970.

Occupational therapy assistants work under the supervision of professional occupational therapists to aid in the rehabilitation of patients who are retarded physically and mentally. Rehabilitational activities are usually carried out through instruction in creative skills, work-related recreational and social functions, or self-care skills. Approximately 6,000 occupational therapy assistants were employed in the United States in 1969, most of whom were women. Most of these assistants qualified through on-the-job training received in hospitals and other health care facilities. Young people seeking careers as occupational therapy assistants should possess good physical and mental health, a sincere liking for people, and the ability to establish and maintain effective personal relationships. Employment opportunities for occupational therapy assistants are expected to be excellent throughout the 1970s. (No refs.) - *B. J. Grylack.*

2423 **CHESS, STELLA.** The child psychiatrist's role in mental retardation. *Journal of Special Education*, 4(2):149-155, 1970.

Psychiatrists should be encouraged to increase their training and involvement in the problems of prevention, therapy, and rehabilitation of MRs. The problems presented by MR are so immense that close collaboration between psychiatrists, psychologists, and educators is needed. When organic disease or behavioral difficulty is suspected, appropriate specialists should be consulted. An adequate differential diagnosis identifies those MR children who require medical attention. A behavioral assessment by a psychiatrist may identify normal behavior, defensive patterns, underlying psychopathology, cerebral dysfunction, or environmental factors. The primary issue involved in the allocation of funds for MR research should be to permit the pursuit of approaches in all disciplines which may be productive. (8 refs.) - *J. K. Wyatt.*

New York University Medical Center
New York, New York

2424 **HARRISON, SAUL I.; McDERMOTT, JOHN F., JR.; SCHRAGER, JULES; & SHOWERMAN, EARL R.** Social status and child psychiatric practice: The influence of the clinician's socioeconomic origin. *American Journal of Psychiatry*, 127(5):652-658, 1970.

The socioeconomic background of the child psychiatrist influences his professional judgment. This conclusion was based on the analysis of data obtained from a questionnaire completed by each clinician studied. The primary comparison was made between clinicians with an upper middle class and those with a lower (or lower middle) class background. There was a tendency for each socioeconomic clinician to assign harsher diagnoses to a child from his own social status. Lower class clinicians reported more aggressiveness and recommended more radical therapy in lower class children. (7 refs.) - *E. Kravitz.*

Children's Psychiatric Hospital
Ann Arbor, Michigan 48104

2425 **BUDNER, STANLEY; ARNOLD, IRENE; & GOODMAN, LAWRENCE.** The plan and the reality: Training and utilization of paraprofessionals for service to the retarded. *American Journal of Public Health*, 61(2):297-307, 1971.

A pilot project undertaken by Retarded Infants Services, Inc., a voluntary agency in New York City, trained low income, unskilled people as aides to professionals in caring for retarded children. All but 2 of the 52 trainees were women, most were minority group members, but none represented the lowest strata of the poverty sector. The training emphasized pragmatic skills that involve performance rather than conceptualization and observation. To overcome the trainees' initial reluctance to participate in group discussions, the first meetings of the 3 groups were filled with specific activities. Lectures were minimized and didactic material was presented in concrete, specific, personal, or dramatic fashion. Audiovisual aids were used heavily. The 3 weeks of practical experience in a retardation program proved more effective when shifted from the end to the middle of the 12-week period so that trainees could review their experiences and thus better relate them to their classroom learning. Important considerations in providing a facilitating environment for training were found to be: (1) financial support during the training period; (2) built-in salient sources of real and symbolic status; (3) avoidance of any possible hints of status derogation of the trainees or of their occupational goal. Further, even "middle class oriented" trainees bear the scars of poverty and discrimination; some attempt should be made to allow the trainees to face up to and resolve these matters. Of the 45 persons who completed training, 73% were employed, 67% in retardation or related areas. Over half the trainees placed had to be provided a temporary salary subsidy as agencies' funds were insufficient for hiring new personnel. Short-term follow-up of job performance indicated that the graduates did very well in terms of motivation, commitment, dedication, and interpersonal skills but less well in terms of initiative and resourcefulness. Despite being trained specifically for work with MRs, many were working effectively with the multiply-handicapped or the blind. (4 refs.) - *J. C. Moody.*

Fordham University School of
Social Service at Lincoln Center
New York, New York 10023

2426 **LINTON, THOMAS E.** A national mental retardation manpower model. *American Journal of Orthopsychiatry*, 42(4):319-320, 1972.

A nationwide Canadian study on emotional and learning disorders in children recommended a generic training model for professionals and paraprofessionals. This developed from the recognition of a serious shortage of trained personnel and the lack of a nationally agreed upon curriculum or training program for MR workers. The National Institute on Mental Retardation set up 4 task forces to study the 4 problem areas of manpower training, preschool and home care, residential care, and occupational-vocational programs, with the focus upon developing manpower and training criteria. Data were collected by surveys, questionnaires, in-depth interviews, library analysis of manpower studies, and analysis of existing manpower training programs. The findings of these task forces resulted in recommendations for nationally coordinated and standardized training models, 4 levels of training for workers in residential nonpublic school settings, and emphasis in manpower training upon developmental handicaps and specialized options. (No refs.) - *S. Glinsky.*

University of Illinois at Chicago Circle
Chicago, Illinois

2427 **LINTON, THOMAS E.** The European Educateur model: An alternative and effective approach to the mental health of children. *Deficience Mentale/Mental Retardation*, 21(2):8-12, 1971.

The European Educateur approach to helping disturbed and retarded children is the suggested basis for a highly innovative program. Reeducation through a program directed by a professional Educateur in this model emphasizes the total approach. The Educateur does not just focus his attention on the child's daily life, but rather he enters into and becomes part of that life. He has both a therapeutic and pedagogic function, and provides a model of personal behavior and concern for a small group of 10-12 children. Extensive use is made of vocational training, physical education, and arts and crafts as a primary means of re-integrating the child into a reluctant social system. Before such a program of helping the child to learn how to establish or re-establish relationships with children and adults can be effective, careful behavioral observation and diagnosis prior to court sentencing or institutional placement are essential. An additional advantage of this model is its ability to directly alleviate the shortage of trained manpower in the mental health field. Educateurs are usually chosen at age 18 and have academic equivalents of a community college degree. The training period usually requires 3 years, with half of this time spent in field work. (No refs.) - *P. Custers.*

No address

2428 **DAVID, CHARLOTTE.** The British Columbia Mental Retardation Institute. *Deficience Mentale/Mental Retardation*, 21(3):2-4, 1971.

The British Columbia Mental Retardation Institute was created to counteract the lack of MR content and experience in professional training. Its aims include demonstration programs to provide models of service in the habilitation of MRs, the training of professional students as specialists in MR, and the promotion of interdisciplinary cooperation among professional workers who are studying and serving the retarded. The techniques used to achieve these aims consist of classes in MR in school departments and clinical experience. The present faculty members at the Institute represent the fields of home economics, nursing, pediatrics, physical education, psychiatry, recreation, social work, and special education. (No refs.) - *B. J. Grylack.*

British Columbia Mental Retardation Institute
British Columbia, Canada

2429 **BALDWIN, WILLIE KATE.** The preparation for certification of teachers for the mentally retarded. *Journal for Special Educators of the Mentally Retarded*, 7(2):93-97, 105, 1971.

Despite apparent significant progress in teacher education and special classes during the past 50 years, many people still have doubts regarding the teachers selected, the training that is offered, the techniques employed by teachers, and the facilities provided for children, especially in the area of MR. One of the main controversial

questions in this area concerns the validity of the requirement in some states that the teacher first secure her elementary or secondary teaching certificate and teach for 2 to 3 years in a regular class with so-called normal children before she is permitted to become a teacher of MRs. It appears that this previous training may not be necessary in the case of teachers who will work with EMR and particularly with TMR children. However, a thorough background in child growth and development and sufficient experience with normal children to allow a normative referent are still considered essential. (No refs.) - *B. J. Grylack.*

Glassboro State College
Glassboro, New Jersey

2430 **BALL, THOMAS S.** A statewide operant conditioning training program for MR hospital employees: The practical results. *California Mental Health Research Digest,* 9(1):10-20, 1971.

A training program in the application of operant conditioning to the treatment of MRs—given at Pacific State Hospital in California for employees of other California State hospitals—seeks to assure commitment of the state government's policy-influencing administrative hierarchy. Initiated in 1968, the first training program in an MR facility was 9 weeks long, 2 of which involved work with nonretarded psychiatric patients. Federal funds have permitted expansion and more formalized operant-conditioning course work. Curriculum and other program aspects are detailed. A cost-benefit analysis indicates that substantial reduction in institutional-care costs results from the behavior-modification programs. (6 refs.) - *B. Berman.*

Pacific State Hospital
California

2431 **HARDY, MADELINE I.** *Standards for Educators of Exceptional Children in Canada.* Toronto, Canada, Council on Exceptional Children, 1971, 173 p. $3.00.

Recommendations as to course content and sequence of training necessary for the adequate preparation of teachers, administrators, and supervisory personnel in the special education field were developed by the Committee on Teacher Education and Professional Standards in Canada. A particular effort is made to locate the role and status of special education within the overall concerns of education generally. Fundamental questions about the nature of MR and the limitations that this disability places on educational possibilities are investigated. The Committee emphasizes that, as is the case in the education of normal children, differences among individual children have as much significance for the teacher as do intra-group similarities. Recommendations for a more integrated approach to teacher preparation in the special education field are developed in specific sections covering models, knowledge, utilization, responsibilities, and accreditation. Appendixes provide additional information on manpower needs in Canada, training center competencies in specific curriculum areas, and existing Canadian courses for special educator training. - *N. Mize.*

2432 **HOLT, K. S.** The training of therapists engaged in the child health and rehabilitation services. *Developmental Medicine and Child Neurology,* 12(5):680-681, 1970. (Letter)

Therapists can make an important contribution to the child health services only if certain positive measures are taken. In the past, occupational, physiotherapeutic, and speech therapists have not been able to contribute fully to child health services due to inadequacy of their undergraduate and postgraduate training, a large, early attrition rate resulting from lack of recognition and poor pay, and a general lack of support of their work. The remedy for this situation involves adequate study of childhood development and allied areas in formal school training, the inclusion of therapists in child health rehabilitation teams, the development of codes of therapeutic practice to achieve satisfactory standards, the informing of therapists regarding the future of their profession, and support and stimulation of their work in every possible way. (No refs.) - *S. Glinsky.*

Wolfson Centre
London WC1N 2AP, England

2433 **WILLIAMS, DAVID M.** An experiment in in-service training: A two-year part-time

course for assistant supervisors at training centres for the severely sub-normal. *Teaching and Training*, 9(1):15-19, 1971.

The Monmouthshire Education Authority's part-time residential course for the staffs of its training centers for the mentally handicapped is based on the recommendations of the Report of the Standing Mental Health Advisory Committee of the Central Health Service Council. The course was operated as a series of residential weekends, one weekend in each month. Practical information was stressed, but the curriculum covered normal and subnormal development, psychology, education principles and practice, medical aspects of MR, etc. Despite overlapping areas of study, sufficient scope was given to the provision for separate development of practical work relevant to Junior and Adult Center staffs. The students' work was assessed in terms of course assignments, a written examination at the end of the 2-year course, and practical teaching ability. A series of sessions in group dynamics and therapy intended to provide a spontaneous welding together of the various aspects of the curriculum into a multisensory approach to curriculum development in the Centers further led to experiments in the use of color in music-making, movement therapy, and developing an audiovisual alphabet by relating sound and color impulses to the initial stages of letter and word recognition. These experiments probably would not have been initiated if it were not for the wide multidisciplinary experiences of the Tutorial and Advisory Panel which designed the program and was largely responsible for its success. (10 refs.) - *J. C. Moody.*

The Hill Residential College
Abergavenny, Monmouthshire, England

2434 **PHILLIPS, MARGARET.** Nutrition manpower in the fields of mental health and mental retardation: The national scene. In: Springer, Ninfa Saturnino, ed. *Proceedings of a Conference on Nutrition and Mental Retardation.* Ann Arbor, Michigan, University of Michigan Publications Distribution Service, 1971, p. 13-22.

There is a serious shortage of manpower to provide the services needed to handle the nutritional problems (screening, diagnosis, followup, etc.) of the retarded. The University Affiliated Training Centers for MR represent an innovative and coordinated approach to the problem. Discussion of this manpower problem entails information on the size of the retarded children population (there are about 6 million retarded persons in the U.S., of whom 2½ million are under 20 years old), the nature of required nutrition services, and available and needed manpower. Nutritional status and skills must be periodically evaluated, the family must be counseled, and the nutritionist must work closely with the entire health team. Adequate nutrients must be available to the fetus and infant. Nutrition manpower is crucial: only 500 positions are available in state, county, and local programs throughout the country. Additional personnel are sorely needed in all areas and at professional, technical, and auxiliary levels. Increased graduate training, study of manpower problems, and better coordination are imperative. (7 refs.) - *B. Berman.*

2435 **LLOYD, ARMESIA.** Nutrition manpower needs in the fields of mental health and mental retardation: On the state level. *Conference on Nutrition and Mental Retardation.* Ann Arbor, Michigan, University of Michigan Publications Distribution Service, 1971, p. 23-24.

The Food Service Section of the Mental Health Department in Michigan has done very little about nutrition, but is now taking steps to make training available within the department and to participate in the University training programs. The Food Service Committee has: developed and evaluated cycle menus; investigated and tested the practicality of prepared entrees; developed a computerized program for menu nutritional values, purchasing, food costs, etc.; and made plans to train all personnel involved in feeding. (No refs.) - *B. Berman.*

2436 **DONNELLY, ERNESTINE.** Interdisciplinary panel presentation: Solving feeding problems of mentally retarded children—Nutritionist. In: Springer, Ninfa Saturnino, ed. *Proceedings of a Conference on Nutrition and Mental Retardation.* Ann Arbor, Michigan, University of Michigan Publications Distribution Service, 1971, p. 77-81.

In a resident institution, feeding the retardate must be organized, not hit-or-miss, since nutritional deprivation increases retardation problems. The nutritionist must use an interdisciplinary approach in solving the problems of feeding the retardate. With the brain-damaged, who have weakened sucking reflexes, gastrostomies with nasogastric tube feeding may be necessary. In such cases, progression and regression may occur for no apparent reason. Consistency of food is important, and it may be necessary to avoid obesity from overfeeding. For different age groups, caloric variations may be necessary. The list of problems is endless and is not solved by institutionalization. The nutritionist must plan, and must work with the nurse, psychologist, speech therapist, and special educator in teaching the retardate to feed himself. Constant training of attendants is necessary in caring for the residents' nutrition. (No refs.) - *B. Berman.*

2437 **HAYS, CAROLE.** Interdisciplinary panel presentation: Solving feeding problems of mentally retarded children—Occupational therapist. In: Springer, Ninfa Saturnino, ed. *Proceedings of a Conference on Nutrition and Mental Retardation.* Ann Arbor, Michigan, University of Michigan Publications Distribution Service, 1971, p. 83-88.

Essential in feeding the retarded child are evaluation of his developmental level, coordination of an interdisciplinary team, and suitable use of a reward system in inducing behavior modification. The approach must be program- and child-centered, not profession-centered: interdisciplinary, not departmental. Depending on the child's readiness, these feeding methods can be used: (1) other than by mouth, if he cannot suck or swallow, with a prescribed formula given by nasogastric or gastrostomy tube; (2) by mouth, if there is a lip reflex and he can swallow his saliva; (3) sucking and swallowing; (4) spoon feeding, if he can open and shut his mouth and swallow a thickened formula; (5) cup feeding; (6) finger foods, if he can munch; (7) cup and a glass. (5 refs.) - *B. Berman.*

2438 **GREEN, LEILA.** Interdisciplinary panel presentation: Solving feeding problems of mentally retarded children—Physical therapist. In: Springer, Ninfa Saturnino, ed. *Proceedings of a Conference on Nutrition and Mental Retardation.* Ann Arbor, Michigan, University of Michigan Publications Distribution Service, 1971, p. 89-96.

The physical therapist must help the retarded child reach his highest possible level of independence. Skill in feeding the multiply handicapped retardate comes with practice and with study of normal and retarded children's actions. Each person working with a patient must be mindful of the whole person. The physical therapist is primarily concerned with the retardate's ability to move in his environment and seeks not only to develop necessary prerequisite skills, but to identify those things that prevent or retard development of skills. The therapist's broad goals include: (1) prevention or alleviation of contractures and deformities; (2) improvement in maintenance of muscle strength, joint-motion range, endurance, and coordination; (3) development of the highest possible level of functional skills; and (4) development of improved breathing patterns. The last is especially for the cerebral palsy victim, the largest single group that physical therapists work with in MR centers. Therapists should instruct parents in how to handle the cerebral palsied child. (No refs.) - *B. Berman.*

2439 **PROVITT, EVELYN.** Interdisciplinary panel presentation: Solving feeding problems of mentally retarded children—Nurse. In: Springer, Ninfa Saturnino, ed. *Proceedings of a Conference on Nutrition and Mental Retardation.* Ann Arbor, Michigan, University of Michigan Publications Distribution Service, 1971, p. 97-99.

The crux of the problem of feeding and maintaining adequate nutritional status in the retarded is prevention, that is, getting the child to eat. Trained personnel are needed; the nurse is very important in an interdisciplinary nutritional program, especially during intensive habilitative training. The nurse must contribute to the interdisciplinary training of personnel, especially in disseminating information on the importance of nutrition to the mother of a retarded child, to those in the community who are taking MR children into their homes, and to all the institutions that care for the retarded. (No refs.) - *B. Berman.*

2440 **SPRINGER, NINFA SATURNINO.** Nutritionists in University Affiliated Centers: A new breed. In: Springer, Ninfa Saturnino, ed. *Proceedings of a Conference on Nutrition and Mental Retardation.* Ann Arbor, Michigan, University of Michigan Publications Distribution Service, 1971, p. 111-123.

A new type of nutritionist has been produced by the University Affiliated Centers, whose development, stemming from a need for professionally trained individuals to undertake research and training in MR, has stimulated broader knowledge and greater understanding through many-faceted interdisciplinary training. The scope of nutritionists thus trained includes training, service, research, administration, and professional development, in addition to clinical duties and consultation. Training activities include teaching formal credit courses and supervising trainees in clinical settings. Topics of research include: amino acid, lipid, and sulfatide disorders; myelomeningocele; obesity; feeding handicapped children; and survey of phenylketonuria. Full-time nutritionists spend much time in professional development, attending seminars, in-service meetings, and staff meetings. The modern nutritionist understands in greater depth the many aspects of her profession. (6 refs.) - *B. Berman.*

AUTHOR INDEX

The author index has been compiled on the basis of abstract numbers rather than pagination.

Aagenaes, Oystein, 1837
Abbas, K. A., 2373, 2378
Abbott, Robert E., 2221
Abramovich, D. R., 1975
Ackerman, Bruce D., 1667
Ackerman, Peggy T., 2179
Adachi, Masazumi, 1873, 1976
Adam, Avinoam, 1960
Adams, B., 1803
Adams, Forrest H., 1921
Aicardi, J., 2008
Ajzenberg, D., 1934
Akesson, H. O., 2374
Akiyama, Y., 1610
Alagille, D., 1657
Al-Aish, Matti S., 2083, 2087
Albee, George W., 1550, 1551, 2333
Alberman, Eva, 1602
Alberty, J., 1938
Alexander, Fred, 1954
Alexander, Linda, 2254
Alexiou, D., 2075
Allderdice, Penelope W., 2041, 2042
Allen, Fred H., 2042
Alley, Gordon R., 2183
Allison, A. C., 1813
Almack, Ronald B., 2330
Altorjay, I., 1939
Al-Ubaidi, Y., 1846
Amatayakul, O., 1653
Amiel-Tison, C., 1919
Ammann, A. J., 1669
Anderson, David E., 1869
Anderson, G. C., 1608
Anderson, Gail V., 1649
Anderson, Gerald G., 1591
Anderson, N. G., 2028
Anderson, R. M., 1977
Anderson, Susan, 2219
Andres, Catherine, 2398
Angeli, Emily, 1641
Angell, Roslyn, 2049
Angenent, H. L. W., 2172
Applegate, Dorothy, 2199
Arakaki, David T., 2070
Archibald, Dorothy, 2299
Arias, Irwin M., 1681
Armstrong, Donald, 1982
Arnold, Irene, 2425
Aronson, Stanley M., 1797
Arora, S. C., 1658
Arto-Medrano, F., 1914
Ashton, A. W., 2193
Ashworth, Maureen, 2029
Atkins, Leonard, 1570, 2037
Aubry, Richard H., 1564
Auerbach, Aaron G., 2216
Auerbach, Victor H., 1607
Austin, James, 1982, 1990, 1995
Axelrod, Saul, 2244
Azanza, X., 1938
Azoury, Ramez S., 1597

Babin, J.-P., 1691
Babra, Mareia de, 2356
Bachhawat, Binual K., 1982
Bachner, Paul, 1738
Baerlocher, Kurt, 1628, 1853
Baker, Bruce L., 2289
Baker, Charles, 2397
Balandin, D., 2240
Baldwin, Willie Kate, 2429
Ball, Thomas S., 2430
Ballabriga, A., 1904
Balsan, S., 1787
Barakat, Bassam Y., 1597
Barbarani, V., 1750
Barcinski, Marcello, 2032
Barclay, D. L., 2031
Barmeyer, George H., 2119
Barness, Lewis A., 1607, 1870
Barnett, Robert, 1615
Baroni, A., 1852
Bartak, Lawrence, 2004
Bartsch, P., 1583
Bartz, Wayne R., 2385
Battin, J., 1937, 1938
Bauer, Karinne, 1736
Bauer, Raymond B., 1683
Bauer, Robert P., 1913
Baumeister, Alfred A., 2157, 2257
Baxt, Roland, 2276
Baylis, Mary, 2020
Beardmore, Thomas D., 1885
Beasley, Nigel, 2194
Behrman, Richard E., 1666
Beischer, Norman A., 2033
Belmonte, M. M., 2043
Bendel, R. P., 2109
Benjamin, Rachel, 1631
Benohr, H. C., 2080
Benson, Philip F., 1825, 2053
Bentovim, A., 1776
Beratis, Nicholas, 2056
Berg, J. M., 2076
Berger, Harriet, 2262
Bergner, Lawrence, 1962
Berk, Paul D., 1689
Berkowitz, Samuel, 2252
Berlin, Nathaniel I., 1689
Berman, Julian L., 1794, 1901
Berry, Helen K., 2144
Best, Gary A., 2295
Beter, Thais R., 2207
Beutler, E., 1843
Bhagat, M., 2242
Bhoyroo, S. K., 1605
Bialac, Verda, 2360
Bianchi, Thomas A., 1906
Bicknell, Joan, 2288
Bieder, J., 2010
Bieri, James, 2131
Bigley, Robert H., 1884
Biglieri, Edward G., 1913
Billings, David M., 2019
Birch, Herbert G., 1805, 2143
Birk, Sharon A., 1643
Birnbaum, Stanley J., 2100
Bischoff, A., 1853
Bissonnette, John M., 1752
Bittner, K., 2074
Blake, Sue, 2253
Blanc, William A., 1759, 1974
Blehova, Bohunka, 1824
Blid, Evy, 2393
Blondheim, S. H., 1719
Bloom, Arthur D., 1800
Bloomer, Joseph R., 1689
Bloomfield, Daniel K., 1916
Blume, K., 1659
Blume, K. G., 2080
Bobath, Berta, 2307
Boda, D., 1939
Bogair, N., 1680
Boley, Scott J., 1887
Bolognese, Ronald J., 1609
Bomsel, F., 1919
Boncompagni, P., 1750
Bonnet, H., 2071, 2072, 2073
Bonte, C., 1920
Borer, Robert C., Jr., 1591, 1608
Borgaonkar, Digamber S., 1629, 2040
Bornstein, Myer S., 1722
Bowen, Trevor, 2335
Bowser-Riley, Frank, 1825
Boyer, Samuel H., 1724
Boylan, Jeanne, 1802
Bracke, P., 1829
Bradford, H. F., 1626
Brady, Roscoe, 1982
Brazelton, T. B., 2187
Breg, W. Roy, 2041, 2042
Bren, G., 1864
Brenchley, Maureen, 1807
Brenner, Paul H., 1591, 1608
Brewer, Fran, 2396
Brink, Joyce D., 1767
Brittain, Harry, 2116
Brobmann, Guenther F., 1672

Brock, D. J. H., 2046
Brody, Irwin A., 1912
Brogger, Anton, 2039
Brookhouse, Diane B., 2137
Brosset, A., 1583
Browder, J. Albert, 2051
Brown, Bertram S., 2338
Brown, Eleanor S., 1910
Brown, James B., 2033
Brown, Jean, 2155
Brown, Richard J., 2349
Brown, Roy I., 2274
Broyer, M., 1787
Brumley, George W., 1758
Brunngraber, Eric G., 1795
Bruns, Paul D., 1752
Bryant, Jean, 2081
Brzeski, Jerzy, 1646
Bucher, Bradley, 2251
Buck, A. R., 1635
Buckingham, Dorothy, 2302
Budner, Stanley, 2425
Budoff, M., 2185
Buhi, William C., 1643
Bundey, Sarah, 1930
Burger, Gary K., 2162
Burgess, Michael M., 1581
Burk, H. Warren, 2370
Burns, Jean, 1778, 1802
Bushnell, Roger K., 2382
Busk, Jytte, 2136
Busse, Walter (Mrs.), 2359
Butler, L. J., 2102, 2110
Buttfield, I. H., 1849
Byles, Thea, 2394
Byrne, Earl B., 1709
Byrnes, Mary E., 1622
Bzostek, Barry, 1996

Cahill, Rose, 1823
Caldwell, Bettye M., 2160
Callahan, Edward W., Jr., 1736
Callander, D. J., 2263, 2316
Callis, L., 1904
Campbell, Barbara, 1838
Campbell, H. Edwin, 2116
Campbell, Stuart, 1578
Carr, Janet, 2138
Carson, Max, 1731
Carter, C. H., 2059
Carter, C. O., 1635
Carter, John L., 2294
Carton, D., 1827
Cartwright, Carol A., 2235
Cartwright, G. Phillip, 2235
Casaer, P., 1610, 1972
Cashdan, Asher, 2415
Cassady, George, 1615, 1956
Castellino, Ronald A., 1924
Castello, F., 1904
Cattaneo, F., 1750
Catti, A., 2104
Cavazos, Ninfa, 1818
Chacko, Claramma M., 1820
Chamberlain, Geoffrey, 1579
Chance, Arthur, 2375
Chang, Alvin C. K., 1672
Channarond, J., 1830
Chase, H. Peter, 1851
Chase, Helen C., 1622
Chatagnon, P.-A., 2164
Chatrian, G. E., 2006, 2007
Chen, Ping Tuan, 1782
Ch'eng, Leslie Y., 2078
Chess, Stella, 2423
Chessells, Judith M., 1740
Chevrie, J. J., 2008
Childers, Ralph L., 1545
Chin, James, 1710
Chin, Tom D. Y., 1745
Chinn, Nina, 1705
Choi, Kyoo W., 1800
Chou, S. M., 1796
Chow, Bacon F., 1858
Chowdhury, N. N. Roy, 1915
Christian, Joe C., 1820, 2040, 2108
Christiansen, Robert O., 1734
Chrysostomidou, O., 2075
Chunga, F., 1693, 1694
Clancy, Helen, 2402
Clark, Sandra H., 1793
Clausen, Johs, 2120, 2315
Clayton, Barbara E., 1776
Cleary, A., 2228
Cleland, Charles C., 2170, 2247
Clemmens, Raymond L., 1762
Cline, David W., 2300
Clot, M., 2399
Clow, C. L., 1850
Coffin, Grange S., 2093
Cohadon, F., 1936
Cohen, Lawrence, 1566
Cohen, M., 1699
Cohen, Michael E., 1946
Cohen, Michael I., 1887
Coleman, Mary, 1883
Coleman, Richard, 2038
Colle, E., 2043
Collins, Edith, 2097
Collins, Hardin A., 2162
Collins, Virginia, 2290
Collipp, P. J., 1864
Collyer, Kathy, 2411
Colwell, David G., Jr., 1553
Conneally, P. Michael, 2040
Connolly, J. H., 1698
Constantopoulos, G., 1874
Cook, William A., 2385
Coop, Richard H., 2137
Cooper, Jack R., 1980
Corson, Stephen L., 1609
Cortazzo, Arnold D., 2343
Corvin, Gerda, 2272
Courtless, Thomas F., 2338
Cowie, Valerie A., 1569
Cox, Alice M., 2406
Cragin, Wesley E., 2207
Cravey, C. Eugene, 1782
Cravioto, Joaquin, 1909
Cremer, N., 1831
Critchley, E. M. R., 2111
Croall, J., 1966, 1967
Crocker, Allen C., 1836
Crome, L., 1989
Crosby, Warren M., 1672
Croughs, W., 2003
Crowley, Jacqueline M., 2020
Crowther, D., 1699
Cruikshank, William H., 1562
Cserhati, E., 1986
Cumming, G. R., 1653, 2103
Cummings, Ann, 2232
Curl, Charles W., 1590
Cuttino, John T., Jr., 1888
Czeizel, A., 1541
Czekalski, S., 2074
Czudner, Gad, 2125

Dabiere, Carol S., 1851
Dacremont, G., 1827
Dalton, Noel T., 1565
D'Amodio, Mabel D., 1889
Damyanov, I., 1949
Dancis, Joseph, 1953
Daniel, William L., 1859
Dargassies, Saint-Anne, 2420
Das, J. P., 2134
Dautrevaux, M., 1784
David, Charlotte, 2428
Davidson, V. Trent, 2336
Davies, Pat, 1554
Davis, Barbara Alphin, 2252
Davis, Bernard D., 1630
Davis, Jessica, 2042
Davis, Larry E., 1745
Davis, Richard J., 2030
Davis, Starkey D., 1799
Davis, William J., 1753
Daw, Edward, 1573
De Angulo, M. S. Laurent, 1900
Deaton, John G., 2418
Debdas, Aloke, 1915
deCapoa, Adriana, 2041, 2042
Dechamps, L., 1659
De George, Frances V., 1620, 1798, 2118
Degnan, Mark, 2019
De Groot, C. J., 1871
Dehan, M., 1919
Dekaban, A. S., 1874
Delacato, Carl H., 1766, 2229
de la Chapelle, A., 2049
Delaney, J. J., 1741
D'Elia, R., 1852
Deligeorgis, D., 2075
Delvin, E., 1850
DeMaegd, M., 1857
DeNardo, Gerald L., 1924
Dennett, X., 1977
Deno, Evelyn, 2215
Denys, P., 1659
De Schrijver, F., 1828
Desjardins, P. D., 1713
Deutsch, Martin R., 2158
Diamond, Louis K., 1631
Diasio, Robert B., 2057
Dietzman, Dale E., 2083
DiGiovanna, Frank, 2330
Dingle, J. T., 1810
Dinniss, Peter E., 2266
Dishotsky, Norman I., 2079
Dobbing, J., 1898
Dodson, W. Edwin, 2083, 2087
Doherty, Daniel, 2162
Dolnick, Michael M., 2281

Dommergues, J.-P., 1919
Donaldson, Lois J., 1724
Donnelly, Ernestine, 2436
Donoghue, Elaine C., 2373, 2378
Doris, John, 1546
Dormoy, O., 1934
Dossa, D., 2071, 2072, 2073
Doxiadis, S. A., 2098
Drash, Allan, 1861
Drazancic, Ante, 1642
Drew, Clifford J., 2135, 2247
Drillien, C. M., 1940
Drolette, Margaret, 2060
Drukker, Bruce H., 1598
Dryman, Irving, 2181
Dubois, Reuben S., 1906
Dubowitz, V., 1917
Duche, D. J., 2366
Dunjic, A., 1659
Dutton, G., 1930
Dutz, W., 1949
Dykman, Roscoe A., 2179
Dyment, Paul G., 1769

Easson, William M., 2000
Ebbin, Allan J., 1710, 2088
Ebrahim, Assma, 1804
Echt, Charles R., 1566
Edsall, Geoffrey, 1631
Edwards, Joseph, 2201
Edwards, R. G., 1632
Edwards, R. Philip, 2183
Eeckels, R., 1829
Eggermont, E., 1829
Ehlers, Kathryn H., 2118
Eid, E. E., 1832
Elbualy, S., 2113
Elchardus, J.-F., 1747, 1919
Eldjarn, Lorentz, 1834
Eller, Eleanor, 2082
Elliott, Raymond N., Jr., 2202
Elwood, J. H., 1945, 1971
Ely, David J., 2124
Emberger, J.-M., 2071, 2072, 2073
Emmanouilides, George C., 1921
Emmery, L., 1829
Empson, Judith, 2030
Engel, Eric, 2030
Engle, Mary Allen, 2118
Ensminger, E. Eugene, 2208
Ente, Gerald, 1664
Ertal, Norman A., 1611
Estela, Luis A., 1743
Eto, Yoshikatu, 1792
Evans, D. E., 1845
Evans, K. A., 1635
Evans, Roy, 1554
Eversden, I. D., 1763
Eyring, Edward J., 1791

Fabia, Jacqueline, 2060
Faidherbe, D., 2010
Fain, Toby, 1543
Faris, Amin A., 1979
Farkova, H., 1978
Farquhar, J. B., 1644
Farquhar, James W., 1842
Farriaux, J. P., 1784
Fasola, Alfred F., 1789
Fedrick, Jean, 2123
Feigin, Ralph D., 1878
Feingold, Murray, 1543, 1587, 2086
Feldmann, Harry, 2184
Fell, J. H., 2284
Fendell, Norman, 2260
Fenichel, Gerald M., 2064
Ferguson, Neil, 1554
Ferguson-Smith, M. A., 2090
Ferguson-Smith, M. E., 2090
Fernandez, Paulina, 2181
Fernandez DeCastro, Aurora, 1687
Fialkow, Philip J., 1780, 2081
Finnie, Nancie R., 2307
Fitzgerald, Joseph A., 2116
Fleckenstein, Loran D., 1979
Flinois, C., 1920
Floquet, J., 1985
Fogel, Bernard J., 1580
Fontaine, G., 1784
Ford, Robin, 1794
Forehand, Rex, 2157, 2257
Forfar, John O., 1712
Forman, Mark, 2391
Forrest, D. M., 2335
Forssman, H., 2374
Fort, Arthur T., 1957
Fortune, Denys W., 2033
Fosker, A. P., 1579
Fox, Irving H., 1885
Fox, Judith, 2282
Fox, Sheila, 2298
Foy, Bob, 2197
Francis, Dorothy E. M. .1776, 1896
Frankel, Jacob J., 1761
Frankenburg, William, 2082
Franks, David J., 2211
Fraser, F. Clarke, 1637, 1961
Fraser, William I., 2161, 2242
Fraser Roberts, J. A., 1635
Freedman, D. G., 2167, 2186, 2187
Freeman, John M., 1770, 2009
Freeman, Roger D., 1547, 1599
Freesemann, C., 1845
Frenk, S., 1846
Fried, Amnon, 1960
Fried, K., 1679
Friedman, J. M., 2031
Friedman, Ronald J., 2212
Friesen, H., 1868
Fuhrmann, W., 2105
Fujiwara, Tetsuro, 1921

Gacs, G., 1986
Gadd, R. L., 2090
Gadow, Enrique C., 1720, 1738, 1963
Gaffney, Thomas E., 1789
Gajdusek, D. Carlton, 1992
Gal, E., 2373, 2378
Galbraith, Gary C., 2136
Gall, G., 1848
Galloway, R. K., 1571
Gambetti, P., 1876
Ganter, Robert L., 2182
Gardner, James M., 2386
Gardner, Leslie, 2177
Gardner, Lytt I., 1943, 1996
Gardner, Riley W., 1768
Gardner, William I., 2250
Garmezy, Norman, 2002
Garrett, Alice L., 1767
Garrett, James F., 2268, 2332
Garrod, O., 2076
Gartler, Stanley M., 1838
Gatmaitan, Zenaida, 1681
Gaudier, B., 1920
Gautier, M., 1657
Gellis, Sydney S., 1543, 1587
George, K. P., 2050
Gerald, Park S., 1886
Gerbie, Albert B., 1594
Gerbie, Melvin V., 1594
Gercken, Gunther, 1670
German, James, 2118
Gersht, N., 2101
Gertner, Melvin, 2032
Gheradi, Gherardo J., 2086
Giannelli, F., 1825, 2049
Gibbs, Clarence, 1992
Gibson, Jack L., 1742
Gigonnet, J.-M., 1933
Giunta, Frank, 1726
Glass, Robert H., 2057
Glenn, Linda, 1735
Gliddon, Jack B., 2136
Gliedman, Marvin L., 1887
Gluck, Louis, 1591, 1608
Goedde, H. W., 2080
Goldfield, Michael, 1690
Goldhamer, Herbert, 2146
Goldschmidt, E., 1835
Goldstein, Harvey, 1602
Goldstein, Herbert, 2236
Goldstein, Joseph L., 1838
Goldstein, Robert, 1592
Gomes, W. J., 1773
Goodman, Harold, 1663
Goodman, Lawrence, 2425
Goodman, Stephen I., 1822, 2051
Goodstein, Henry A., 2178
Gordon, J. G., 2293
Gordon, Neil, 2016
Gorlin, Robert J., 1950
Gortuny, G., 1904
Gottlieb, Jacques S., 2001
Graham, William E., 2116
Gray, Clinton W., 1724
Grech, P., 1966, 1967
Green, Elizabeth A., 2287
Green, J. R., 2006, 2007
Green, Leila, 2438
Greenberg, Robert E., 1734
Gregson, N. M., 1568, 1575
Griffis, Bertram W., 2332
Griffith, John F., 1983
Grignon, G., 1985
Groffman, Helen, 1955
Gropp, A., 2105
Gross, Howard A., 1649
Gross, Jerry C., 2128, 2176
Grosshans, E., 1933
Grossman, Frances K., 2413
Guenter, Kenneth E., 1743
Guerrin, F., 1920

Guess, Doug, 2208
Guillard, J.-M., 1830, 1936
Guiter, F., 1830
Gulbis, E., 1659
Gunzburg, H. C., 2364
Gusdon, John P., 1673
Guthrie, Richard, 1735

Haberland, Catherine, 1795
Habib, R., 1787
Hadden, Diana S. M., 1674, 1698
Haddow, J. E., 1848
Haffter, C., 1596
Haicken, Barry N., 1911
Haire, Margaret, 1674, 1698
Hakim, C. A., 1572
Halbrecht, I., 2101
Hale, William R., 1767
Hall, Sylvia M., 2170, 2203
Hall, W. Knowlton, 1782
Halliday, N. P., 1603
Hallock, James A., 1870
Hamanaka, Toshihiko, 1981
Hambert, G., 2063
Hambidge, K. Michael, 1906
Hammel, Deborah, 1607
Hammer, N., 2366
Hanefeld, F., 1989
Hanrahan, Harriet, 2233
Hansen, J. D. L., 1845
Hanson, Harley M., 1858
Hanson, Robert M., 2262
Hansson, O., 2026
Harcke, Howard T., 1759
Hardisty, R. M., 2089
Hardley, P., 2388
Hardy, Madeline I., 2431
Harper, Rosemary, 2228
Harries, J. T., 1896
Harris, Jerome S., 2095
Harris, Maureen, 1639
Harris, R., 2090
Harris, Ruth F., 1897
Harrison, Saul I., 2424
Harshman, Fred, 2354
Harvin, D., 1699
Hashem, Nemat, 1804
Haworth, J. C., 1653
Hayes, M. F., 1658
Hays, Carole, 2437
Haywood, Scott, 2255
Heaton-Ward, W. Alan, 2362
Hecht, Frederick, 2081
Heffner, R. R., Jr., 1984
Heffner, Reid R., 1968
Hegarty, John, 2194
Hehunstre, J.-P., 1691
Heiman, Melvin L., 1649
Held, Berel, 1711
Hellegers, Andre E., 1752
Hellman, Louis M., 1611
Henderson, Norman D., 2148
Henle, Gertrude, 1716
Henneman, Carol E., 1649
Hennig, Heinz, 2188
Henri, Jacques, 2270
Henton, Karel R., 2258
Herbertson, Leon M., 1769
Herbst, Bernadette A., 1946
Herndon, Robert M., 1770
Herschkowitz, N. N., 1988
Hertzig, Margaret E., 2143, 2181
Hetzel, B. S., 1849
Hide, T. A. H., 1763
Higgins, James V., 1859
Hill, Hoyle D., Jr., 1888
Hill, Leighton L., 1847
Hinkel, G. K., 1695
Hirschhorn, Kurt, 2032, 2035, 2056
Hirth, L., 2080
Hirvonen, T., 1660
Hjalmarsson, O., 1833
Ho, Oi-Lan, 2112
Hogan, Michael A., 2116
Hohenauer, Leonhard, 1889
Holden, Kenton R., 1954
Holden, Raymond H., 1866
Holliday, Audrey R., 1553
Holliday, Malcolm A., 1913
Hollowell, Joseph G., 1782
Holmes, Lewis B., 1925, 2037
Holt, K. S., 2432
Holt, Laurence E., 2128, 2176
Hommes, F. A., 1871
Hong, R., 1669
Hook, Ernest B., 1633
Hopkins, M. S., 1687
Hopps, Hope E., 1753
Horky, Karel, 1717
Horstmann, Dorothy M., 1685, 1730
Horton, W. A., 1865
Houillon, P., 2010
Howat, J. M., 1964
Hsu, Lillian Y. F., 2032, 2035, 2056
Hug, George, 1884
Humbel, R., 1985
Hunt, J. McV., 2339
Hunter, Janet L. P., 1773

Ihsan, Al-Issa, 2231
Ikeda, Takayoshi, 2094
Ingram, Irene, 2411
Ingram, Robert A., 2175
Isacson, Peter, 1744
Isler, W., 1853
Itokawa, Yoshinori, 1980
Iwanska, Janina, 1646

Jackson, James L., 2269
Jacobson, Cecil B., 2048
Jagenburg, R., 1833
James, G. B., 1662
Jancar, J., 2067
Jans, J., 1970
Jarman, Ruth V., 1707
Jarvis, Jane A., 2085, 2096
Jassani, M., 1584
Jean, R., 2071, 2072, 2073
Jeffree, D. M., 2415
Jenkins, David M., 1644
Jenkins, G. H. C., 1604
Jenner, F. A., 1812
Jennings, A. N., 2240
Jensen, N., 1774
Jepson, Anne M., 2341
Jepson, John B., 1809
Jervis, George A., 2322
Jewelewicz, Raphael, 1892
Johnson, Betty Jo, 1553
Johnson, Charles F., 2084
Johnson, G. Frank, 1950
Johnson, Gil, 2277
Johnson, K. E., 2200
Johnson, Kenneth P., 1682
Johnson, Lois, 1656
Johnson, Margaret Anne, 2410
Joiner, Lee M., 2128, 2176
Jones, G., 1699
Jones, Howard W., Jr., 1597
Jones, Mary, 2306
Jones, R. N., 2055
Jones, Walter S., 1866
Jongbloet, P. H., 2058
Jordan, J., 1584
Josey, Julian C., 1598
Juchau, Mont R., 1589
Julien, Claude, 2271
Junker, Karin Stensland, 2393
Justice, R. S., 2345
Justice, Wanda P., 2218

Kadotani, Tetsuji, 2094
Kahn, Lawrence I., 1878
Kahn, Max, 2350
Kaiser, H., 1840
Kaiser, I. H., 1721
Kajii, Tadashi, 1943
Kallen, David J., 1855
Kalverboer, A. F., 1972, 2166
Kamaryt, J., 2061
Kang, Ellen Song, 1886
Kapitulnik, J., 1719
Kappelman, Murray M., 2182
Karambula, Katerina, 1821
Karczewski, Mieczyslaw, 2323
Kardon, Nataline, 2056
Karnes, Merle B., 2196
Karon, Myron, 1736
Karrer, Rathe, 2120
Kattamis, Christos, 1821
Katz, Elias, 1563, 2412
Katz, Samuel L., 1733, 1983
Katz, Selig, 1802
Kaufman, Robert L., 1878
Kehrer, Anthony F., 1744
Kelley, Vincent C., 1860
Kelley, William N., 1885, 1893
Kelly, Charles S., 1742
Kelly, Sally, 1778, 1802
Kemball, M. L., 1652
Kempe, C. Henry, 1764
Kenny, Thomas J., 1762
Kersey, J. H., 2109
Kershner, John R., 2405
Kesmodel, Kristin, 2340
Keys Smith, G., 2241
Khalili, Abbass, 1557
Kijima, Shigeji, 1981
King, Larry W., 2251
King, Saddie, 1943
Kint, J.-A., 1827
Kintzel, H.-W., 1695
Kirman, Brian, 1641, 1645, 1931

Kitagawa, Teruo, 1972
Klaus, E., 1978
Klein, R., 1848
Klein, Sarah, 1856
Kletzky, O., 2101
Klinger, Harold P., 2042
Klumb, David (Mrs.), 2359
Kneebone, G. M., 1894
Knowles, B. A., 2239
Knox, E. G., 1544
Kobayashi, Mitsuo, 1611
Kodanaz, Altan, 1581
Koehl, R. H., 1984
Koenig, Ronald H., 1973
Kogon, Alfred, 1856
Kohn, Gertrude, 1582
Kohorn, E. I., 1685
Komlos, L., 2101
Kornfeld, M., 1875
Korniszewski, L., 2066
Korns, Robert, 1778
Kouznetsov, V. N., 1785
Kowal, Areta, 2118
Krasner, Joseph, 1589
Krmpotic, Eva, 2099
Krugman, Saul, 2376
Kubarych, Sandra F., 1738
Kucera, Jiri, 2091
Kuhl, W., 1843
Kulovich, Marie V., 1591, 1608
Kupfer, C., 2007
Kushnick, Theodore, 1542

Labrune, B., 1747
Lacey, Stephen H., 1791
Lahr, Irene W., 2297
Lake, Brian D., 1771
Lakoff, Kenneth M., 1609
Lampert, Peter W., 1992
Lane, Pauline M., 2214
Lappin, T. R. J., 1731
Lardinois, R., 1694
Larroche, J.-C., 1919
Larson, H. Elliott, 1753
Larson, Thalia, 2245
Laski, Bernard, 1965
Lattin, John E., 1769
Laxova, Renata, 2061
Leake, Norman H., 1673
Lebenthal, Emanuel, 1960
Le Coultre, R., 1972
Lee, C. L. Y., 1568, 1575
Lee, J. C., 2109
Lee, John C. M., 1997
Lee, T. K., 1713
Lee, Tzu Y., 1567
Lehrnbecher, Wolfgang, 2048
Leibovitz, Albert, 1742
Leicht, Kenneth L., 2152
Leman, P., 1936
Lenneberg, Eric H., 2171
Lennette, Edwin H., 1710
Lequien, P., 1920
Lerique-Koechlin, A., 1935, 2013
Lerner, A. Martin, 1683
Leroy, Jules G., 1826, 1844
Lesch, Michael, 1772
Lessey, Robert, 1890
Lettich, E., 2006, 2007
Leumann, Ernst P., 1913
Levi, A. Jonathan, 1681
Levine, Edna S., 2190
Levison, Cathryn A., 1998
Levitan, Max, 1882
Levy, E. P., 1961
Levy, Harvey L., 1877, 1902
Lewak, Norman, 1732
Lewis, Kay, 1951
Libo, Shirley S., 2299
Lie, Sverre O., 1834, 1837
Liebhaber, H., 1685
Liebig, Margaret, 2389
Lim, Henry S., 1752
Limbeck, George A., 1860
Lindenbaum, R. H., 2102
Lindsay, Gavin, 1842
Lines, David R., 1819
Linsao, L., 1755
Linton, Thomas E., 2426, 2427
Lipscomb, Wendell R., 2079
Lister, A. H., 1918
Littlefield, John W., 1570
Lloyd, Armesia, 2435
Lobb, Harold, 2121
Loeb, H., 1831
Loening, W. E. K., 1783
Loggie, Jennifer M. H., 1789
Lohmann, Werner, 2126
Lohr, G., 1659
Lohr, G. W., 2080
Loken, Aagot Chr., 1837
Long, Robert E., 1588
Lopes, Antonio C., 1720
Lotker, L., 2101
Lotkin, Pauline, 1879
Lott, Ira T., 1902
Loughman, William D., 2079
Lowry, R. B., 1862
Loy, Donald L., 2385
Lozzio, Carmen, 1879
Lucas, Alexander R., 2001
Lucas, George J., 2048
Lucas, Marilyn S., 2132
Luck, Elizabeth, 2182
Lugo, Gustavo, 1956
Lundberg, P. O., 2047
Luse, Sarah A., 1890
Lyagoubi, Souad, 2018
Lyon, G., 1872

Macafee, Jeremy, 2033
Mace, John W., 1822
Maciejewski, J., 2074
Mackay, C. K., 2155
MacKay, D. N., 2291
MacKenzie, G., 1945
Mackenzie, S., 1850
MacMahon, Brian, 1926
MacMillan, D. C., 1965
MacMillan, Donald L., 2213
MacQueen, John C., 2212
Madden, David L., 2083
Madeley, B. R., 2318
Madigan, Phyllis M., 1877
Madoff, Morton A., 1631
Mahon, D. F., 1544
Malin, A., 2265
Malkova, E. V., 1785
Mallarive, J., 1936
Mallet, R., 1747
Maloney, Michael P., 2173
Mamula, Richard A., 2417
Man, Evelyn B., 1866
Maney, Ann C., 2369
Mantle, D. J., 2089
Marchal, C., 1985
Marchand, J.-Cl., 1937
Marcuse, Edgar K., 1705
Marek, Josef, 1717
Marienfeld, Carl J., 1557
Marini, A., 1750
Marks, V., 2020
Marshall, John, 2024
Marshall, Linda D., 1588
Marshall, Tony, 2313
Martin, C., 1691
Martin, E. A., 1891
Martin, Madelyn H., 2195
Martin-Bouyer, G., 1919
Martinez, S. Alicia, 1701
Martinov, Y. S., 1785
Mathers, N. P., 1662
Mathieson, Gordon, 1770
Matoth, Yehuda, 1761, 1960
Matsaniotis, Nicolas, 1821
Matsuoka, Tatsunori, 1981
Matthew, Earl B., 2083
Matthews, Geraldine M., 2357
Mattinson, Janet, 2169
Maxwell, Jane E., 2301
Mayo, O., 1777
McCall, Robert B., 2127
McCauley, John J., 2222
McCreary, Sally A., 1643
McDermott, John F., Jr., 2424
McDonald, Ian, 2256
McDonald, John A., 1893
McDonald, T. P., 1879
McEnery, G., 1654
McGee, Barbara J., 2030
McGilvray, Eve, 2042
McIlwain, Henry, 1627
McIver, C., 1652
McKusick, Victor A., 1880, 1950
McMullin, G. P., 1658
Mehta, Arvind C., 1903
Meile, Richard L., 2370
Melin, Bengt, 2285
Mellman, W. J., 1737
Mendenhall, Hiram W., 1618
Menkes, J. H., 1977
Menolascino, Frank J., 2296
Merlis, J. K., 2012
Merrill, Robert E., 1556
Metaxotou-Mavromati, Anna, 1821
Metcoff, J., 1846
Meyer, Harry M., Jr., 1708, 1753
Meyer, John Sterling, 1683
Michie, Eileen A., 1634
Migeon, Barbara Ruben, 1788
Milhorat, Thomas H., 1923
Milic, Ann M. B., 1974
Miller, Alfred P., 2276
Miller, Dorothy A., 2042
Miller, Gerald L., 1745

Miller, Harold R., 2258
Miller, L. H., 1701, 2006, 2007
Miller, Marion C., 1842
Miller, Orlando J., 2041, 2042
Miller, Phillis K., 2294
Millian, Stephen J., 1856
Mills, Millie, 2240
Milner, R. D. G., 1652
Milstein, Victor, 1576
Milunsky, Aubrey, 1570
Minchin, A. J., 1605
Minkowski, A., 1919
Mir, G. H., 2103
Mirkin, Bernard L., 1704
Mises, J., 1935, 2013
Mitchell, B. Jean, 1888
Mitra, Sudhansu B., 2392
Moffatt, W. R., 2291
Mogar, Robert E., 2079
Mohammed, Shakil, 1789
Mohyuddin, Fazl, 1628
Monch, E., 1840
Monif, Gilles R. G., 1618, 1711
Monkus, Ellen St. J., 1580
Monset-Couchard, M., 1919
Moodie, A. D., 1845
Moosa, A., 1917, 1994
Morales, M., 1846
Mordock, John B., 1756
Morgan, Paul N., 1707
Morris, C. Richard, 1847
Morrison, Donald F., 2369
Morrow, Grant, III, 1607, 1870
Moskowitz, Herbert, 2126
Muhlethaler, J. P., 1555
Muir, Helen, 1811
Muller, Helga, 2027
Muller, Paul F., 2116
Muller, Victor H., 2116
Muranyi, L., 1939
Murdock, A. I., 1755
Murken, J.-D., 2105
Murphree, Oddist D., 1707
Murphy, Ann, 1586
Murphy, D. A., 2043
Murray, M., 2246, 2267
Myant, M. B., 1814
Myrianthopoulos, Ntinos C., 1797, 2139, 2150
Myszkowski, Leopold, 1646

Nachtigall, Lila E., 1892
Nadler, Henry L., 1594, 1820, 1843, 1884
Naeye, Richard L., 1759, 1974
Naggan, Lechaim, 1942
Nagyfy, Staphan F., 1943
Naik, S. N., 1869
Najenson, Theodore, 1960
Nakao, T., 1748
Nance, Walter E., 2040, 2108
Navis, Ronald (Mrs.), 2359
Navon, Ruth, 1908
Naylor, Alfred F., 1797, 2139, 2150
Naylor, Harriet, 2348
Neeman, Renate L., 2130
Nellhaus, Gerhard, 1614
Nelson, Calvin C., 2209
Nelson, Matilda M., 1712
Nelson, Thomas L., 1548, 1799
Nesbitt, Robert E. L., Jr., 1564
Neu, Richard L., 1943, 1996
Neufeld, Elizabeth F., 1781
Neville, B. G. R., 1771, 1897
Nezelof, Christian, 1692
Nickel, Vernon L., 1767
Nirje, Bengt, 2324, 2363
Nishikawa, Tadashi, 1981
Niswander, Kenneth R., 1589, 1749
Nitzberg, Jerome, 2273
Nitzen, Menachem, 1955
Nocton, F., 1657
Nolan, David C., 1683
Noland, Robert L., 2414
Nour, Assma, 1804
Nourse, C. H., 1652
Noyes, Andrea N., 1724
Nuechterlein, Keith, 2002
Nyhan, William L., 1508, 1931

Oakey, R. E., 1644
Obaldia, Mario de, 2295
O'Brien, Donough, 1851
O'Brien, John S., 1790, 1844, 1905
O'Connell, Edward J., Jr., 1588
O'Connor, Gail, 2345
O'Connor, William A., 2419
Odievre, M., 1657
O'Donoghue, S. E. Fidelma, 1723
O'Grady, Donald J., 2144
Oh, William, 1889
O'Halloran, Mary T., 1803, 1895
Ohama, Koso, 2094
O'Harra, Edwin A., 2264
Oka, Eiji, 1574
Okada, Shintaro, 1790, 1844
Okken, A., 1871
Okubovejo, A., 1944
Olanders, S., 2063
Oldendorf, William H., 1903
Oldfield, R. K., 1894
Olney, John W., 2112
O'Neill, Robert T., 1607
Ornitz, Edward M., 1997
Ortiz, Eduardo, 1598
Osborne, Robert G., 2296
Osofsky, Howard J., 1588
Ostendorff, Martha E., 1782

Pace, Robert, 2369
Packham, D., 2228
Padeh, Baruch, 1908
Paget, G. E., 1751
Pak, Yong Cha, 1760
Palkes, Helen, 2180
Pallisgaard, G., 1835
Palmer, Carolyn, 2299
Pank, Penelope, 2115
Pantelakis, S. N., 2098
Papiernik, Martine, 1692
Parkman, Paul D., 1708, 1753
Pashayan, H., 1961
Passarge, Eberhard, 2080, 2118
Patel, Daksha A., 1666
Patry, George, 2018
Patton, Mary E., 2258
Paulus, Dieter H., 2189
Pazoutova, Noemi, 1824
Pedersen, Jens Malling, 2361
Peneva, Petrana, 1877
Penfold, J. L., 1894
Pennington, J. A., 2205
Perkins, Kay, 2305
Peters, John E., 2179
Peterson, Donald R., 1705
Petry, Judith J., 1633
Pfeiffer, Rudolf A., 1950
Pharoah, P. O. D., 1849
Phibbs, R. H., 1699
Phillips, Herbert E., 2130
Phillips, Margaret, 2434
Phillips, Robert, 2262
Picciano, Dante, 2048
Pickel, F. W., 2223
Pickering, Douglas, 1965
Pickersgill, Mary J., 2115
Pierson, M., 1786
Piesowicz, A. T., 1896
Pildes, Rosita S., 1663, 1666
Pincus, Jonathan H., 1980
Pine, Nellie H., 2305
Piret, L., 1659
Piros, Katalin, 1980
Pitt, D., 2015
Plato, Chris C., 2087
Plotkin, S. A., 1737
Pluss, H. J., 1853
Polani, P. E., 2049, 2050
Polansky, Norman A., 2409
Pollak, Shlomo, 1822
Pollitt, Ernesto, 2145
Pollitt, R. J., 1812
Pomeroy, Diana, 2368
Pond, D. A., 2023
Ponte, C., 1920
Pope, Michael, 2292
Portal-Foster, C. W., 1560
Porto, Sergio O., 1663
Posner, Bernard, 2279
Posternak, Yvonne, 2353, 2367, 2383
Pounds, Lois, 1586
Pounds, V. A., 2011
Power, R. M. H., 1713
Pozsonyi, J., 1868, 2092
Preece, J. M., 2020
Prehm, Herbert J., 2135, 2204
Prentice, Norman M., 2131
Prichard, Robert W., 1673
Primrose, D. A., 1922
Pritchard, Jack A., 1714
Proesmans, W., 1829
Provitt, Evelyn, 2439
Puck, Mary, 2082
Punter, Agnes, 2234
Purdy, J. M., 1632
Pustel, Gabriel, 2158
Pye, Carole, 2089

Queenan, John T., 1720, 1738, 1963
Quinlan, D. C., 1891

Race, R. R., 2049
Rahal, James J., Jr., 1716
Raiha, Niels, 1921
Raine, D. N., 1808, 1815
Rampini, S., 1853
Rapp, Fred, 1686
Ratner, Gerald, 2129
Ravaris, C., 2140
Rawlings, Grace, 1947
Reddi, O. S., 2068
Reed, Terry E., 2040, 2108
Rees, H., 2055
Regius, Mary, 2028
Reid, Duncan E., 1619
Reid, M. McC., 1755
Reid, Winifred M., 1675
Reifsnyder, David N., 1701
Reiss, H. E., 2110
Remy, J., 1920
Renuart, Adhemar W., 1950
Renwick, D. H. G., 1862
Renzulli, Joseph S., 2189
Revesz, C., 1541
Rey, J., 2071, 2072, 2073
Reynolds, E. O. R., 1757, 1947
Reynolds, John W., 1760
Reynolds, William A., 1598
Rezvani, I., 1864
Rhyne, A. Leonard, 1673
Rhys, Ann, 1890
Ribierre, M., 1747
Riccardi, Vincent M., 2037
Richards, B. W., 2034, 2065
Richards, Sharon F., 1600
Richardson, Alan, 1763
Richardson, B., 2384
Richens, Alan, 2021
Ridler, M. A. C., 2076
Rieu, D., 2071, 2072, 2073
Rimbot, B., 2013
Rimoin, David L., 2036
Rittenhouse, A. H., 2116
Ritter, Linda, 1656
Ritvo, Edward R., 1997
Rivera-Olmos, Victor M., 1683
Robertson, Jean, 2304
Robertson, John G., 1634
Robine, Gerald G., 2235
Robinow, Meinhard, 1950
Robinson, Arthur, 1582, 2082, 2095
Roboz, P., 2015
Rodgerson, Denis O., 1906
Rodjer, S., 1833
Roeder, Lois M., 1858
Rohlfing, Barbara, 1951
Romeo, Giovanni, 1788
Rondini, G., 1697
Roopnarinesingh, Syam, 1606
Rosado, A., 1846
Rose, Vera, 1965
Rosemberg, S., 1872
Rosenthal, Bernard S., 2261
Rosnick, Michael R., 2099
Ross, Robert T., 2153
Rossi, E. E., 1988
Roszkowski, Irenaeusz, 1646
Roth, J., 1669
Rothschild, Edmund O., 1727
Rourke, Byron F., 2125
Rourke, R. J., 2395
Routh, Donald K., 2404
Roux, J. F., 1584
Royer, P., 1787
Ruben, R. J., 1729
Rubinstein, Lucien J., 1770
Rudiger, H. W., 1659, 2080
Rundle, A. T., 2122
Runnels, Eugene J., 2343
Rusalem, Herbert, 2276
Rutter, Michael, 2004
Ryan, G. B., 1977
Ryan, Joanna, 2168
Ryback, David, 2237

Sabatino, David A., 2340
Sacrez, R., 1933
Saint-Jammes, Tortes, 1936
Salmon, Michael A., 2029
Sandbank, Uriel, 1960
Sandler, Louise, 2129
Sanger, Ruth, 2049
San Juan, B., 1691
Saudubray, J.-M., 1919
Saugstad, Letten F., 1779
Sayre, James W., 1715
Scarr, Sandra, 2210
Schaefer, Edward W., Jr., 1724
Schedel, Ingolf, 1670
Schenker, J. G., 1719
Scher, Barry M., 1881
Schiff, D., 1652
Schiffer, Morton A., 1611
Schimke, R. Neil, 1865
Schindeler, Joan, 2059, 2113
Schlenker, Janet, 1982
Schmalhofer, Alicja, 1646
Schmid, Rudi, 1736
Schmid, W., 2104
Schmidt, Leo J., 2209
Schneck, Larry, 1873, 1976
Schoenbaum, Stephen C., 1651
Schongut, L., 1986
Schrager, Jules, 2424
Schroter, Werner, 1670
Schruefer, John J. P., 1752
Schubert, William K., 1789
Schuler, D., 1986
Schulte, F. J., 1612
Schutta, Henry, 1656
Schwarz, Richard H., 1870
Schwarze, R., 1695
Schwinger, E., 2105
Scott, C. Ronald, 1793
Scott, Daniel E., 1714
Scott, H. Denman, 1709
Scott, Ronald C., 1838
Scriver, Charles R., 1628, 1850
Seakins, J. W. T., 1896
Seeds, A. Elmore, 1752
Seegmiller, J. Edwin, 1816
Seelenfreund, Moshe, 1960
Seham, Max, 1999
Serafica, Felicisima C., 2142
Sergovich, F., 2092
Sever, John L., 2083
Severi, Francesca, 1697
Shaffer, Thomas E., 1791
Shah, Saleem A., 1629
Shapiro, Lawrence R., 2032, 2035, 2069
Sharlin, Shlomo A., 2409
Sharpley, Robert P., 1553
Shear, Carol, 1931, 2113
Shennan, Victoria, 2286
Shepard, Frank M., 1929
Shepard, Thomas H., 1739, 1799
Shepherd, Jean, 1776
Shepherd, Jocelyn, 2243
Sheridan, Edward P., 2407
Sheridan, Kathleen, 2407
Sherry, Patrick J., 2252
Shochet, Solly Barnard, 1960
Showerman, Earl R., 2424
Shuster, Arnold A., 2217
Shuster, Susan K., 2217
Shuto, T., 2062
Siddiqui, A. R., 2291
Siegel, Bess, 2407
Siegel, Paul S., 2156
Siegler, J., 1986
Silberberg, Donald H., 1656
Silver, Larry B., 2114
Silverman, Joseph A., 1703
Simons, Christine, 2086
Simonson, Maria, 1858
Simpson, P. F., 2314
Singer, J. D., 2046
Singleton, Edward B., 1951
Sisson, Warden B., 1903
Siva Sankar, D. V., 2106
Skaberne, Bronislav, 2165
Skjei, Priscilla, 2421
Skrede, Sverre, 1834
Slater, Nathan M., 2282
Slater, Stanley, 1595
Slaton, Paul E., 1913
Slovis, Thomas L., 1906
Smith, Alfred A., 1953
Smith, Ellen M., 1553
Smith, G. F., 2059, 2113
Smith, James O., 2208
Smithurst, B., 2028
Snider, Bill, 2183
Sofuni, Toshio, 2094
Sokal, G., 1659
Solitare, G. B., 1984
Sollee, Natalie D., 1886
Solomon, J. B., 1684
Solowy, Marian K., 1929
Sommers, Paul, 2128, 2176
Song, A. Y., 2283
Song, R. H., 2283
Soper, Harry H., 2351
Sosa, A., 1846
Sparkes, Robert S., 1838, 2027
Spellacy, William N., 1591, 1608, 1643
Spencer, D. A., 2377, 2379
Sperling, Mark, 1861
Spirer, Z., 1680
Spiro, Alfred J., 1973
Spranger, Jurgen W., 1826, 1863
Springer, Ninfa Saturnino, 1561, 2440
Stachowiak, James, 2419
Stafford, Calvin, 2129

Staub, Henry P., 1665
Stavlenic, Ana, 1642
Steele, Mark W., 2041
Stefan, H., 1840
Stenchever, Morton A., 2085, 2096
Stephen, Elspeth, 2304
Stephens, Rosemary, 1771
Steptoe, P. C., 1632
Stern, Carolyn, 2201
Stern, Diana J., 1897
Stern, H., 1654
Stern, Jan, 1897, 1931, 2052
Sternberg, B., 1935
Sternberg, W. H., 2031
Sternlicht, Manny, 2158
Stevens, Janice R., 1576
Stewart, Ann, 1947
Stewart, Mark, 2180
Stiavelli, R. E., 2223
Stimmler, L., 1774
Stinnett, Raymond D., 2204
Stoebner, P., 1933
Stoelinga, G. B. A., 2151
Stokke, Oddvar, 1834
Stoll, C., 1933
Stone, Nellie D., 2334
Strande, Carl, 1703
Strang, L. B., 1947
Stratford, Bernice F., 1932
Strauss, Ronald G., 1789
Streissguth, Ann Pytkowicz, 1739
Stromme, Johan H., 1834, 1837
Stuckey, S. J., 1895
Stumpf, David, 1982, 1990, 1995
Subrt, I., 1824
Summer, George K., 1888
Summitt, Robert L., 1950, 2107
Susser, Mervyn W., 1962
Sutcliffe, R. G., 1777
Sutherland, Betty S., 2144
Sutton, M. D., 1755
Suzuki, Kunihiko, 1836, 1982, 1987, 1991, 1993
Suzuki, Yoshiyuki, 1836, 1987, 1991
Swartz, Jon D., 2247
Swedberg, Kathryn R., 1793
Swinyard, Chester A., 1648
Swyer, P. R., 1755
Sylvester, P. E., 1823, 2020, 2122
Szczepski, O., 2074

Taii, Shunzo, 1981
Tait, Charles, 1592
Takei, Hiroshi, 2044
Talbot, C., 1699
Talkington, Larry W., 2170, 2203
Tanguay, Peter E., 1997
Tanner, John, 2352
Tapia, Hugo R., 1818
Tassinari, C. Alberto, 2018
Tatebayashi, Yoshinori, 1981
Taylor, David C., 2025
Taylor, Rodney H., 2287
Tejavej, Anant, 1649
Teng, Cecilia Chiang, 1793
Teoh, Eng Soon, 1643
Teplitz, R., 1843
Tettenborn, U., 2105
Thaler, M. Michael, 1678, 1736
Thevaos, Theo G., 1782
Thieffry, St., 1872
Thier, Samuel O., 1817
Thomas, Alexander, 2181
Thomas, Elizabeth C., 2230
Thompson, Hartwell G., 1796
Thompson, Robert S., 1705
Thuline, Horace C., 2081
Thurmon, Theodore F., 1724
Tiernan, J. R., 1652
Tilak, K. S., 1755
Timmermans, J., 1829
Timmons, G. D., 1682
Timson, J., 2090
Tinnefeld, W., 2105
Toivanen, P., 1660
Tomkiewicz, S., 2366
Tondeur, M., 1831
Tooley, W. H., 1699
Torii, Junzo, 1976
Toseland, P., 1774
Touchette, Paul E., 2154
Touchstone, Joseph C., 1609
Touwen, B. C. L., 1765
Treciokas, Leo, 1903
Trembley, Phillip W., 2224
Trinidad, F., 1843
Turner, Brian, 2097
Turner, Gillian, 2097
Turton, Lawrence J., 2308
Tuttle, Dean, 2277
Tweed, Gerald V., 1745

Uilenberg, Catharina, 2092
Ulan, O. A., 1755
Ulrich, J., 1853
Umansky, Irving, 1631
Urbanek, K., 1978
Urrusti, J., 1846
Usategui-Gomez, Magdelana, 1687

Vaitukaitis, J., 1848
Valaes, R. N., 2098
Valderrama, Elsa, 2032
Valenti, C., 1873
Valletutti, Peter, 2163
Vallo, A., 1904
Vamos-Hurwitz, E., 1831
VanCampen, Jack, 2129
Vancon, Nancy, 2401
Van Den Daele, Leland, 2149
Van Der Scheren-Lodeweyckx, M., 1829
Vanderveer, Beverly B., 1739
Van Der Werff Ten Bosch, J. J., 2151
Van Gemund, J. J., 1900
Van Gils, J. F., 1969
Van Hoof, P., 1829
Van Leeuwen, G., 1735
Van Nevel, C., 1857
Van Sande, Marc, 1775
Van Uden, A. M. J., 2117
Variela, M., 2031
Veath, M. Lois, 1790, 1844
Vegni, Maria, 1697
Velasco, L., 1846
Veress, I., 1939
Verger, P., 1830, 1936
Vessey, M. P., 2089
Vidailhet, C., 1985
Vidailhet, M., 1985
Vidyasagar, D., 1746
Viggiani, James C., 2226
Vinik, A. I., 1783
Viukari, N. M. A., 2017
Vlachos, I., 2075
Vlietinck, R., 1827
Volk, B. W., 1873, 1976
Von Premerstein, Richard, 2303
Vrablik, George R., 1724

Wadsworth, H. G., 2416
Wadsworth, Joanna B., 2416
Waechter, Eugenia H., 2403
Wahlstrom, J., 1583, 2047
Waisman, Harry A., 1819, 1910
Walberg, Herbert J., 2147
Walchner, Andreas M., 1878
Walczak, M., 2074
Waligora, A., 2074
Walker, Geoffrey F., 2335
Walker, John, 1647, 1662
Walker, S., 1568, 1575
Walker, W., 1650
Wallace, Helen M., 2344
Wallace, Sheila J., 1655
Waller, H. D., 2080
Walsh, S., 2028
Walter, Donald O., 1549
Walti, Ulrich, 2191
Walz, Donald V., 2078
Warburton, Dorothy, 2041, 2042
Ward, A., 1846
Ward, L. Charles, 2156
Ward, Michael H., 2289
Ward, Michael P., 2173
Warkany, Josef, 1616
Warner, Kenneth, 2001
Warren, Neil, 2345
Warren, Richard J., 2036
Watts, Anne P., 2331
Waxman, Sorrell H., 2070
Wear, Douglas J., 1686
Weaver, L. A., 2140
Weber, A., 1659
Weber, Felice M., 2027
Wei, Ping Y., 1567
Weinburg, M. M., 2278
Weiner, Lawrence H., 2249
Weinstein, Morton R., 1690
Weismar, Richard, 2129
Weiss, Edward S., 1557
Welch, J. Philip, 1841
Welch, N. Noreen, 1851
Wellby, M., 1894
Wells, R. S., 1930
West, Jenny, 2400
Westall, R. G., 1823
Whalley, Peggy J., 1714
Wheelden, Julie A., 1902
White, Philip L., 1854
Whitfield, C. R., 1731
Whittaker, Camilla, 2220
Widdowson, E. M., 1899

Wiecek, Zygmunt, 2022
Wiesmann, Ulrich, 1781, 1988
Wigglesworth, J. S., 1740
Wiglesworth, F. W., 2043
Wilkins, Robert H., 1912
Willerman, Lee, 2139, 2141, 2150
Williams, Barbara L., 1600
Williams, Curtis S., 1742
Williams, David M., 2433
Williams, Jerry D., 2107
Williams, Mair, 2192
Williams, Margaret L., 2210
Williams, Norwood L., 2340
Williams, Phillip, 1554
Williams, Robert L., 1847
Williams, Sandra, 1744
Willoughby, H. W., 1713
Wilson, Arthur L., 2408
Wilson, David J., 1715
Wilson, Hazel, 1744
Wilson, Miriam G., 1710, 2088, 2093
Wilson, R., 1584
Winick, Myron, 1952
Winters, Robert W., 1703
Wolff, O. H., 1776, 1896
Wolff-Plodowska, Anna, 2066
Wollen, W., 2159
Wolraich, Mark, 1996
Wood, C. B. S., 1613
Woo-Sam, James, 1767, 2174
Worth, Dorothy J., 1631
Wright, Harley T., 1557
Wright, Logan, 2133
Wyne, Marvin D., 2137, 2421

Yaffe, Sumner J., 1589
Yamamoto, Kaoru, 2230
Yanagisawa, S., 2062
Yankow, Sanford, 1580
Yap, Lucy L., 1889
Yatsu, Frank M., 1907
Yen, Stella, 1926
Yeni-Komshian, H., 1584
Yeung, C. Y., 1668
Yielding, K. Lemone, 2054
Yoshida, P., 1846
Yoshida, T., 1846
Yu, J. S., 1803, 1895
Yu, Pao-lo, 2040
Yunis, J. J., 2109

Zachary, R. B., 1959
Zahtz, Hyman, 1867
Zaizov, Rina, 1761
Zajaczkowska, Krystyna, 2066
Zamet, P., 1693, 1919
Zang, K. D., 2045
Zankl, H., 2045
Zarfas, D. E., 2365
Zatz, Leslie M., 1924
Zausmer, Elizabeth, 1725
Zaverio, Silvio, 1697
Zealley, Helen, 1655
Zerolis, Joanne B., 2346
Ziegler, Dewey K., 1581
Zlotnick, A., 1679
Zollar, Lowell M., 2099

SUBJECT INDEX

The subject index has been compiled on the basis of abstract numbers rather than pagination.

Abnormalities
fetal, induced by environmental agents, 1638
three-legged anencephalic child, 1944

Abnormalities, Drug Induced, 1712
by coumarin derivatives, 1916
by diphenylhydantoin, 1704
effect of anticonvulsant drug on chromosomes, 2039
LSD and genetic damage, 2079
mutagenic effect of ^{131}I in mice, 2068
neonatal hypoglycemia from antidiabetic drugs, 1652

Abnormalities, Multiple
acrodysostosis, 1950
associated with alaninuria, 1774
associated with C group autosomal mosaicism, 2037
associated with purine metabolism, 1772
Bartter's syndrome, 1789, 1848
cervico-oculo-acoustic dysplasia, 1922
in children; prognostic study, 1556
in Cockayne's syndrome, 1917
congenital encephalopathy and familial dyschromia, 1933
in Cornelia de Lange syndrome, 1859, 1931, 1961
in crying cat syndrome, 2041
with group B (4-5) long arm deletion, 2102
in Hurler's syndrome, 1807
identification of new syndromes in, 1616
kinky hair syndrome, 2019
little leopard syndrome, 1965
Lowe's syndrome, 1835
monitoring for congenital malformations, 1624
in mucopolysaccharidosis, type 1, 1831
in partial C (6-12) trisomy, 2109
in Prader-Labhardt-Willi syndrome, 1937, 1938, 2076
recommendations for dealing with, in France, 1559
Rubinstein-Taybi syndrome, 1951
Swiss study of congenital malformations, 1555
in trisomy C, 2071
in 4p-syndrome, 2042

Abortion
cytogenetic determinations in spontaneous, 2070
spontaneous, in Rh immunization, 1720

Acid-Base Equilibrium
in fetal distress, 1915
in infants born with umbilical cord around the neck, 1914

Acidosis
effect on unbound bilirubin concentration, 1694
fetal congenital lactic, in two siblings, 1834, 1837
metabolic, in infantile diarrhea; tissue hypoxia and defective renal acid excretion in, 1847
renal tubular, therapy for, 1904
tromethamine and sodium bicarbonate in, 1750

Acrodysostosis: See under **Bone and Bones**

Adrenal Glands
in hyaline membrane disease, 1759, 1760

Affective Disturbances
emotional deprivation; influence on growth and behavior, 2003

Alanine
alaninuria, associated with multiple abnormalities and MR, 1774

Allergies
cerebral, mistaken for MR or emotional disturbances, 1754

Amaurotic Familial Idiocy
GM_1 gangliosidosis, 1836
intracytoplasmic lipid bodies in Tay-Sachs disease, 1873
Kuf's disease, 1796, 1875
Tay-Sachs disease, 1797, 1814, 1907, 1908

Amino Acids
abnormal excretion of, in Hartnup's disease, 1809
in amniotic fluid, diagnostic significance of, 1607
blood brain barrier penetration by, and gamma emission of selenomethionine Se 75, 1903
in blood serum of parents of children with congenital malformations, 1646
and brain damage in mice, 2112
content in hair, and inborn errors of metabolism, 1775
defects in renal tubular reabsorption of, 1808
distribution and metabolism of in brain tissues, 1627
membrane transport of, in MR, 1628

Amniocentesis
in genetic counseling, 1594
optimum time for performing, 1583
in United Kingdom, 1569

Amniotic Fluid
antibody titers, in erythroblastosis, 1673
biliverdin in, 1589
contaminated with metheme pigments, in fetal erythroblastosis, 1719
detecting chromosomal abnormalities in, 2110
detection of glycogenosis in, 1884
diagnostic significance of amino acids in, 1607
effect of tromethamine or bicarbonate infusions on, 1752
estriol concentration in, as index of fetal state, 1609
fetal factors influencing volume and composition of, 1975
fetal sex determined by examining cells of, 1597
immunoglobulin G levels and titers of specific antiviral antibodies in, 1618
immunoglobulins in, 1590
as index of fetal lung maturity, 1608
measuring contents of, to determine fetal age, 1649
optical density, as index of fetal outcome, 1565

Amniotic Fluid (continued)
polyploidy in, 1575
proteins, in normal and Rh-sensitized pregnancies, 1738
recovery of cytomegalovirus from, 1745
significance of tetraploidy in, 1570
spectrophotometric analysis of, 1615, 1749
technique for eliminating red blood cells from, 1568
tested, for glycogen storage diseases, 1593
tetraploid cells in, 1582

Anemia
congenital hemolytic, 1659

Anencephalus, 1928
epidemiology of, 1941
epidemiology of, in Boston and Rhode Island, 1926
epidemiology of, in Britain, 1945, 1971
epidemiology of, in Central Virginia, 1929
hydramnios in, 1974
in Israel, 1942
and maternal amniotic fluid, 1567
role of ethnic background in, 1949
role of fetal swallowing in polyhydramnios, 1975

Antibodies
rhesus, as index of fetal outcome, 1565
rubella neutralizing, 1856

Antigen-Antibody Reactions
rubella antibody determination among pregnant women, 1743

Antigens
Australian, incidence in mongolism, 2083
Xg specificity of, 1777

Arthrogryposis
congenital, with MR, 1960

Ascites
urinary, with spina bifida, 1964

Ataxia
walker for child with, 2297

Ataxia Telangiectasia
lymphoblastic transformation, chromosome pattern and delayed type skin reaction in, 1986

Atropine
in mongolism, 2103

Attitude
of EMR, toward significant people and curriculum, 2230
toward MR in Sweden, 2324
in MR, measured by semantic differential, 2242
of parents toward EMR, 2416
of potential employers toward MR, 2279
of public toward MR, and future developments, 1542

Australia
classification and description of mental deficiency cases in, 2015
sheltered workshop for MR in, 2259
trisomy 18 in, 2028

Austria
special education for MR at Levana Institute in Vienna, 2303

Autism (See also Schizophrenia, Childhood)
admission of autistic children to MR hospitals, 2377
attitude of helping professions towards, 2243
behavior therapy in, 2286
and chromosome abnormalities, 1996
chromosome breakage in infantile, 2106
in England, 2198
group holiday workshop for families with autistic children, 2402
music therapy in, 2295
neurobiological abnormalities in, 1997
possible etiologies of, 2004
symptomatic, in childhood and adolescence, 2000

Autonomic Nervous System
activity at rest of, in normals and MR, 2120
dysautonomia, familial, type II and congenital sensory neuropathy, 1953

Bartter's Syndrome: See under **Abnormalities, Multiple**

Battered Child Syndrome
pediatric implications of, 1764

Behavior
and abnormal development of brain, 2151
antisocial, in MR, 2338
characteristics of communicating and noncommunicating MR, 2170
in children with minimum cerebral dysfunction, 1756
effect of emotional deprivation on, 2003
as index of nutritional inadequacy, in rats, 1858
indicative of object concept attainment, in deviant children, 2142
influence of genetics on, 2167
oral language, in normal and EMR children, 2178
stereotyped; head banging in rhesus monkey, 1998

Behavior Therapy
adopted at Patton State Hospital, California, 2305
for autism, 2286
basic concepts and techniques, 2256
concepts and practices applied to MR, 2250
in de Lange syndrome, 1931
for dyslexia, 2237
in education of MR, 2239
in EMR special education class, 2233, 2253
increase in nonadaptive behavior of SMR with nonreward, 2257
for mongoloid boy, 2255
with multiply handicapped child, 2254
pay as incentive for MR work behavior, 2264
for PMR subject, 2258
punishment of nonverbal psychotic child, 2251
and sight, sound and symbol kit to develop basic reading skills, 2232
for SMR, 2289
to develop self-feeding skills in PMR, 2252
token economy program for MR, 2248
training in, for hospital employees, 2430

Belgium
classification of MR in, 1558
financial support for MR in, 2325
incidence of congenital rubella in, 1718
needs of PMR in, 2326

Bilirubin
biliverdin in amniotic fluid in early pregnancy, 1589
conjugation, in newborn rats, 1678
controlled by phototherapy, in neonatal jaundice, 1726
decreasing levels of in premature infants, with orotic acid, 1695
effect of photodegradation products of on cerebellum cultures, 1656
levels in neonatal jaundice, 1691
metabolism, proteins Y and Z in, 1689
unbound, relationship to total bilirubin level and to kernicterus, 1693
unbound, separation by gel filtration and micro-determination of, 1694

Biological Transport
amino acid membrane transport in MR, 1628
tubular, and amino aciduria, 1808

Blood
donors screened for glucose-6-phosphate dehydrogenase deficiency hemoglobin variants, and serum haptoglobin types, 1869
fetal, estimation of pH of, 1571
leukocyte metabolism in intrauterine malnutrition, 1846
lymphoblastic transformation in ataxia telangiectasia, 1986
plasma factor VIII concentrations in XXX women, 2089
Rh negative, protein concentrations in, 1687

Blood Cells
oxygen affinity of, in intrauterine transfusion, 1662
phospholipids in, comparative study, 1670

Blood Platelet Disorders
chronic thrombocytopenia purpura and minimal brain dysfunction, 1761

Blood Proteins
amino acid concentrations in blood serum of parents of children with congenital malformations, 1646

Blood Transfusion
feto-maternal, as index of placental damage and fetal risk, 1605

Blood Transfusion, Intrauterine
action line method for management of rhesus isoimmunization, 1731
and fetal death, in baboons, 1672
followup study of children who received, 1699
oxygen affinity of red blood cells in, 1662

Bone and Bones
acrodysostosis, 1950
pycnodysostosis, 1791
skeletal malformations in 49,XXXXX constitution, 2092

Brain
amino acid uptake studied by measurement of radioisotope content, 1903
and behavior, abnormal development of, 2151
development and malnutrition, in guinea pig, 1851
development, effect of malnutrition on, 1898
maturation and neonatal EEG, 1612
pathology in fetus with Tay-Sachs disease, 1873
weight, correlated with skull circumference size in infant, 2123

Brain Damage, Chronic
art therapy in, 2294
behavioral aspects, 2166
Doman method in, 2298
and dyslexia, 1766
minimal brain dysfunction, 1756, 1762, 2175, 2183
minimal brain dysfunction with chronic thrombocytopenia purpura, 1761
multivariant prediction in diagnosis of, 1581
music therapy in, 2295
neurological indications of minimal brain dysfunction, 1765
neuropsychiatric aspects, 2023
orthomolecular therapy in, 2309
reaction time study in, 2125
value of EEG in evaluating, 1769

Brain Diseases
lipofuscin storage disease, 1977
subacute necrotizing encephalomyelopathy, 1979, 1980
subacute spongiform encephalopathy, 1981

Brain Injury, Acute
impact of on development of cognitive-affective structures, 1768
intellectual function in children following, 1767

British Columbia
Hunter and Hurler syndromes in, 1862

California
behavior therapy at state hospital, 2305
rehabilitation for multiply handicapped blind students, 2277
training hospital employees in behavior therapy in, 2430

Canada
Bevan Lodge, Courtenay, B. C., 2382
British Columbia Mental Retardation Institute, 2428
citizen sponsored MR residential facility in, 2380
Community Mental Health Services in Ontario, 2351
education for the TMR in, 2227
epidemiological study of MR in Prince Edward population, 1560
group home program for MR in New Brunswick, 2352
Regional Mental Retardation Program (Saskatchewan), 2354
residential facilities for MR in, 2365
standards for educators of exceptional children in, 2431
voluntary services for MR in, 2353

Carbohydrate Metabolism, Inborn Errors
new pattern of excretion in, 1823

Carbohydrates
excretion of, in galactosemia, 1782
metabolism of, during intrauterine growth retardation in rats, 1889

Cataract
with hyperphosphatasia and MR, 1773

Cell Membrane Permeability
in Bartter's syndrome, 1848

Central Nervous System Diseases (See also specific diseases)
Creutzfeldt-Jakob disease, 1981, 1992

Cerebellum
cultures, effect of bilirubin photodegradation products on, 1656

Cerebral Aqueduct
stenosis of, with hydrocephalus, following mumps encephalitis, 1682

Cerebral Ischemia, Transient
device for monitoring pressure in, 1763

Cerebral Palsy
description of cases, 2015
parent-staff communication in treatment and management of, 2307

Cerebral Sclerosis, Diffuse
experimental globoid cells produced in rats, 1993
Krabbe's globoid cell leukodystrophy, 1907, 1976, 1982, 1987, 1989, 1991, 1994
metachromatic leukodystrophy, 1985, 1995
metachromatic leukodystrophy and Cockayne's syndrome, 1917
metachromatic leukodystrophy in fibroblasts, enzyme replacement treatment for, 1988
metachromatic leukodystrophy, sulfatase activity in, 1990
metachromatic leukodystrophy with mucopolysaccharidosis, 1853

Cerebrospinal Fluid
dynamics in hydrocephalus, 1923, 1924
immunoglobulin G, in children, 1614
metabolism of, in hydrocephalus, 1927

Chediak-Higashi Syndrome, 1813

Child
autistic, group family holiday for, 2402
behavior of, correlated with neurological manifestations, 2166
with congenital rubella, mental development of, 1739
delinquent, Slovenian study of, 2165
development and psychosocial deprivation, 2160
disturbed; electroencephalograph readings of, 1576
epilepsy in: etiology, epidemiology, and prognosis, 2008
hyperkinetic, intellectual ability and perceptual-motor performance in, 2180
lead poisoning in; medical aspects, 1706
low birth weight and disadvantaged; effect of intervention on performances in, 2210
nonverbal; sign language for, 2245
normal development of applied to guidance of handicapped children, 2304

Child (continued)
parameters of growth in failure to thrive, 1832
psychiatry for, influence of socioeconomic background of therapist on, 2424
rare form of mucopolysaccharidosis in, 1872

Child Development
related to perinatal incidents, 2116

Chromosome Abnormalities (See also **Chromosomes; Sex Chromosome Abnormalities; Trisomy**)
acrocentric supernumerary, 2048
afollicular ovarian dysgenesis and XY chromosome, 2094
associated with C group autosomal mosaicism, 2037
in autism, 1996
autosomal anomalies, 2107
breakage, in infantile autism, 2106
in crying cat syndrome, 2046, 2078, 2104
detection of, in amniotic fluid, 2110
epidemiology of, 1621
extra posterior cervical skin as indicator of, 2035
G deletion syndromes, 2036
Klinefelter's syndrome, 2038
monosomy 21 with mosaic 45,XX,21-46,XX,21 pi, 2072
multiple abnormalities with abnormal D group chromosome, 2059
mutagenic effects of radioiodine in mice, 2068
parental, and multiple abnormalities in offspring, 2032
parental, and reproductive failure, 2096
partial trisomy 13, 2099
partial 5 deletion, and normal triosephosphate isomerase activity, 2080
and plasma growth hormone levels, 2047
in Prader-Willi syndrome, 2076
quintuple X syndrome, 2066
from radiation, physical factors, viruses, and drugs, 2064
in spontaneous abortions, 2070
syndrome fluctuations and month of birth, 2058
in Turner's syndrome, 2027
twinning, and thyroid diseases, 1798
in Wolf-Hirschhorn syndrome, 2093
X, incidence of in neonatal population, 2095
X, prognostic study of neonates with, 2082
Xg locus in structurally abnormal X chromosomes, 2049
X-linked MR, 2097
XX/XO sex chromosomes with MR and deafness, 2067
XXX women, and plasma-factor-VIII concentrations, 2089
XYY males, 2077
Y chromosome in sperm of XYY man, 2057
47,XYY males; meiosis and testicular histology, 2105
49,XXXXX, 2092

Chromosomes (See also **Chromosome Abnormalities; Sex Chromosome Abnormalities**)
changes of, in higher plants, 2055
effect of anticonvulsant drug on, 2039
effect of diphenylhydantoin on, 2085
effect of LSD on, 2079
loss of, and hypodiploidy, 2045
mutations in, by chemicals and drugs, 1639
pattern of, in ataxia telangiectasia, 1986
redundancy of, and gene expression, 2054

Classification
of epilepsies, 2024
of MR, in Belgium, 1558

Cockayne's Syndrome: See under **Abnormalities, Multiple**

Cognition (See also **Intelligence**)
cognitive style and conceptual tempo in MR, 2137
defect of, in autism, 2004
development of, in phenylketonuric children, 2144
development of, related to season of birth, 1554
encoding processes of MR and normals, 2152
impact of brain injury on development of cognitive-affective structures, 1768

Communication
and noncommunication behaviors among MR, 2170

Community Health Services (See also **Community Mental Health Services**)
counseling for MR, 2421
for handicapped, in England, 2347
HEW programs for MR, 2311
hostels for MR, 2375
information center for handicapped children, 1543
learning disability clinic, 2182
for MR, evaluation of, 2344
parent and child centers for disadvantaged, 2339
Regional Mental Retardation Program, Saskatchewan, Canada, 2354
return of aged MRs to community, 2345
Syndrome Identification and Consultation Service, 1587
versus residential facilities treatment, in England, 2378

Community Mental Health Services (See also **Community Health Services**)
group home program in New Brunswick, Canada, 2352
for MR; recommendations for expansion, 2329
in Ontario, Canada, 2351
volunteers in, 2348

Convulsions
febrile, associated with viruses, 1655
neonatal, as indicator of disease, 2009

Coumarins
and fetal deformities, 1916

Counseling
genetic, 1637, 2099
genetic, amniocentesis in, 1594
genetic, effect on parents of MR, 2418
genetic, etiology of MR and, 1641
genetic, in England, 1635
genetic, in rehabilitation medicine, 1648
genetic, in Scandinavia, 1596
parents of MR, 2414

Creatinine
concentrations in amniotic fluid, in normal and abnormal pregnancies, 1606

Cretinism
endemic; iodine injections in, 1849

Creutzfeldt-Jacob Disease: See under **Central Nervous System Diseases**

Crime
patterns of MR in relation to treatment, 2161

Crying Cat Syndrome, 2107
in adolescents and adults, 2041
chromosome abnormalities in, 2078, 2104
contrasted with 4p-syndrome, 2042
in partial chromosome 5 deletion, 2080
triosephosphate isomerase levels in, 2046

Cystinosis
with rickets and renal tubular acidosis, 1904

Cystinuria
and cerebral metabolism, 1817

Cytomegalic Inclusion Disease
intrahepatic cholestasis in, 1657
in microcephalics with MR, 1747

Cytomegaloviruses
atypical cases, 1654
recovery of, from amniotic fluid, 1745

Deafness (See also **Hearing Disorders**)
congenital, and disturbed psychomotor development, 2117
manual sign language programming for deaf MR, 2203
with microphthalmia and MR, 1925
program planning for inst MR children with, 2392
testing for intelligence in, 2190

De Lange Syndrome: See under **Abnormalities, Multiple**

Delivery of Health Care
generic training model for MR professionals and paraprofessionals, 2426
for MR, 2327

Denmark
residential facilities for MR in, 2361

Dermatoglyphics
in diagnosis, 1601
in diagnosis of mongolism, 2040, 2108

Diabetes Mellitus
following hypoglycemia of infancy, 1861
in Prader-Willi-Labhardt syndrome, 1938
in pregnancy, free fatty acid level in, 1642

Diabetic Coma
in case of mongolism, 2043

Diagnosis, 1987
of a new mucopolysaccharidosis, 1863, 1864, 1865
amniocentesis in United Kingdom, 1569
amniotic fluid in genetic counseling, 1594
at-risk register, in England, 1602
of atypical cytomegalovirus infections, 1654
Bender-Gestalt test and differential classification of MR, 2173
of brain damage in cerebrovascular accident population, 1581
of chromosome abnormalities in embryo, 1632
clinical determination of fetal hemoglobin in vaginal bleeding, 1579
dermatoglyphics in, 1601

Diagnosis (continued)
EEG patterns associated with various kinds of MR, 1574
estimation of pH of fetal blood, 1571
extra posterior cervical skin as indicator of chromosome abnormalities, 2035
of fetal distress, by hormone relationships, 1564
of fetal erythroblastosis, by chemical tests of metheme-contaminated amniotic fluid, 1719
fetal monitoring techniques, 1600
of fetal state, by plasma estriol measurements and ultrasonography, 1611
of fetus at risk, by analysis of urinary estriol secretion, 1644
of Gaucher's disease, 1843
of GM1 gangliosidosis, 1836
of GM1 gangliosidosis, type 2, 1827
of hyaline membrane disease, 1591, 1758
of hydrocephalus with dementia, 1958
of hyperuricemia, by enzyme assay, 1580
identification of new syndromes in cases of multiple abnormalities, 1616
immunoglobulins in, 1613
isotopic placentography in placental localization, 1598
of lead poisoning, 1706, 1727
of Lesch-Nyhan syndrome, 1801, 1838
metacarpal sign in ovarian agenesis, pseudohypoparathyroidism and pseudopseudohypoparathyroidism, 1595
of methylmalonic acidemia, prenatally, 1870
of minimal brain dysfunction, 1762, 2175
of mongolism, by dermatoglyphics, 2040
of mongolism, by X-ray, 2100
of MR, by Brunet-Lezine development test, 2191
of MR, by copy drawing test and tree drawing test, 2188
in neonatal seizures, 2009
optical density and amniotic titer predictions, 1565
of phenylketonuria, 1878
physician assessment vs. at-risk register, 1604
and prognosis in phenylketonuria, 1901
of pyridoxine deficiency by tryptophan-tolerance test, 2026
of Refsum's syndrome, 1891
repeat evaluations of MR children, 1586
of rubella, during pregnancy, 1676
of sphingolipidoses, 1815
of subacute necrotizing encephalomyelopathy, 1979
Syndrome Identification and Consultation Service, 1587
of Tay-Sachs disease by hexaminidase activity, 1908
test for glycogen storage diseases, 1593
ultrasonics in monitoring fetal development, 1585
value of EEG in evaluating minimal brain dysfunction, 1769

Diphenylhydantoin
effect on chromosomes of human leukocytes, 2085
placental transfer and neonatal elimination of, 1704

Down's Syndrome: See Mongolism

Drug Therapy
effect of trimethadione and paramethadione on fetus, 2118
neuleptil for epileptic MR, 2022
phenobarbitone in hyperbilirubinemia, 1702
for rickets and renal tubular acidosis, 1904
sulthiame for disturbed MR, 2291
teratogenic effects of, 1712

Drugs
effect of tromethamine or bicarbonate infusions on fetal circulation and amniotic fluid, 1752
metabolization of, by feto-maternal system, 1723

Dwarfism
diastrophic, with rubella, 1665

Dyslexia
behavior therapy for, 2237
and brain injury, 1766
neurological etiology for reading disabilities, 2229

Ecuador
first school for MR in, 2356

Edema
of mother, placenta, and fetus, 1721

Education, Special
administrative needs, 2314
arithmetic as training in analytical listening, 2222
art for slow learners, 2292
art therapy for minimally brain damaged, 2294
for autistic, in England, 2198
behavior therapy in, 2233, 2239, 2253, 2256
British Columbia Mental Retardation Institute, 2428
computerized course to help teachers identify children with learning problems, 2235
developing MR learning capacity in, 2200
for development of functional intelligence, 2224
disadvantaged MR and, 2216
for educable and trainable MR, 2336
the educational psychologist and MR, 2205
ethnic distribution of children in EMR and LD classes, 2211
European educateur model, 2427
evaluating efficacy of, 2209
factors in verbal and motor learning of TMR, 2231
films on MR, 2219
first school for MR in Ecuador, 2356
Frostig program for visual perception development, 2223
habilitative training for social behavior, 2247
for handicapped in England, 2328
Hebrew Academy for Special Children, 2350
in-class work-study program for MR, 2264
individualized performance profiles for MR in, 2214
language programs for disadvantaged, 2201
the library as teaching environment, 2220
of low birth weight disadvantaged children, short term program, 2210
meaningful material in, 2202
for MR, at Levana Institute in Vienna, Austria, 2303
for MR, in Minnesota, 2342
for MR, in Poland, 2323
for MR, in U.S. and abroad, 2236
for MR, their parents, and teachers, 2206
music, dance, and storytelling for TMR and EMR, 2225
music therapy for autistic and brain damaged children, 2295
nature study for SMRs, 2394
for physically handicapped; psychoeducative considerations, 2212
population characteristics of MR schools in Washington State, 2372

Education, Special (continued)
predicting educability of cerebral palsied children, 2177
for preschool MR, 2218
Project UNIQUE for EMR (Rochester, New York), 2226
questionnaire for parents of EMR receiving, 2416
reconstructed and reinforced reality concepts in, 2249
remedial basic education and vocational goals, 2282
requirements and goals, 2193
research and development needs, 2217
and research priorities for MR, 2315
residential courses for MR training center staffs, 2433
standards for educators in, in Canada, 2431
in Sweden, 2316
teaching aids for MR, 2221
teaching art to MR children, 2306
for the TMR, in Ontario, Canada, 2227
token reinforcement programs, 2244
using television to stimulate acquisition of reading skills, 2234
vs. integration of MR into regular classes, 2213
10-point reading program for EMR, 2199

Electroencephalography
EEG readings in children with psychiatric disorders, 1576

Electronics, Medical
electronic monitoring of labor, 1584

Encephalitis
in hamsters, after injection of human measles virus, 1686

Encephalitis Viruses
herpesvirus hominis idoxuridine in, 1683
mumps, followed by aqueductal stenosis and hydrocephalus, 1682

Endometrium
chromosome abnormalities in cells of, 2070

England (See also **Great Britain**)
administration of education for MR in, 2314
assessment of community services for handicapped in, 2347
at-risk register in, 1602
bedroom groupings of handicapped in, 2368
Eastbourne conference on MR, 2319
education for autistic in, 2198
education for handicapped in, 2328
evaluation of demand for short- or long-term care of MR in, 2373
genetic counseling in, 1635
hospitalization of MR in, 2378
infant at risk registers in, 1544
intensive-care program for very low birth weight infants in, 1948
legal status of MR in, 2313
marriages between MR in, 2169
mass vaccination for rubella in, 1751
mistreatment of MR patients in residential facilities in, 2381
residential courses for MR training center staffs in, 2433
residential facilities for MR in, 2362, 2379
short-term hospital care for MR in, 2388

Environment
effect of, on fetus, 1970

Enzymes
activity of, in Cornelia de Lange syndrome, 1859
adenosine triphosphatase inactivation in epileptic seizures, 2017
alpha-galactosidase, in Fabry's disease, 1788
amylase heterogeneity in mongolism, 2061
arylsulfatase A in metachromatic fibroblastic leukodystrophy, 1988
arylsulfatase A in different forms of metachromatic leukodystrophy, 1995
assays of, in diagnosing sphingolipidoses, 1815
beta-galactosidases in Hurler and Sanfilippo syndromes, 1825
beta-glucosidase activity in, in Gaucher's disease, 1843
deiodinase deficiency in hypothyroidism, 1857
disorders of, in ganglioside storage disease, 1814
galactocerebroside beta-galactosidase, 1982
galactocerebroside beta-galactosidase deficiency in Krabbe leukodystrophy, 1987, 1991
galactose-1-phosphate uridyl transferase, 1782, 1820
gamma-glutamyl transpeptidase deficiency and MR, 1822
glucose-6-phosphate dehydrogenase deficiency, 1777, 1821, 1869
hexosaminidase A, in juvenile GM2 gangliosidosis, 1790
hexosaminidase levels in Tay-Sachs disease, 1908
hypoxanthine guanine phosphoribosyltransferase in Lesch-Nyhan syndrome, 1772, 1780, 1800, 1838, 1893
in inborn errors of metabolism, 1806
inhibition of, in subacute necrotizing encephalomyelopathy, 1980
lysosomal, in aspartylglycosaminuria, 1812
red-cell triosephosphate isomerase in crying cat syndrome, 2046, 2080
sarcosina dehydrogenase, in sarcosinemia, 1793

Epidemiology
of anencephalus and spina bifida, 1941
of anencephalus, in Belfast and Scotland, 1945
of chromosome abnormalities, 1621
of CNS malformations, in Hungary, 1541
method for estimating number of cogenital malformations, 1557
of minor congenital malformations unassociated with major congenital malformations, 1633
of MR, in Canadian community, 1560
suicide attempts among MR in institutionalized population, 2158

Epilepsy
associated with natural hypoacusis and heterochromia iridis, 2010
in children, 2008
classification of, 2012, 2024
Comprehension and Picture Arrangements scores of epileptics, 2174
description of cases, 2015
folic acid and vitamin B12 in, 2016
folic acid treatment in, 2021
group therapy for self-induced seizures in, 2299
intelligence and learning in, 2025
and MR, 2011
neuropsychiatric, 2023
pattern sensitive, 2006, 2007

Epilepsy (continued)
post-traumatic, in children, 2013
potassium aberrations in, 2017
sectioning of corpus callosum in, 2014
serum-phenytoin levels in folic acid treatment of, 2020
subclinical electrical status in 6 MR children, 2018
use of neuleptil in cases of, 2022
wigs to replace protective helmets for epileptics, 2302

Erythroblastosis, Fetal (See also Hyperbilirubinemia)
diagnosis of, by chemical test of metheme-contaminated amniotic fluid, 1719
intrauterine blood transfusion in; followup study, 1699
Ovenstone Factor in assessing, 1722
prognostic parameters in, 1673
protein values in, 1738
Rh immunity globulin donor program in Massachusetts, 1631
Rh, phospholipids in blood cells in, 1670
rhesus hemolytic disease, 1658
spectrophotometric analysis of amniotic fluid in, 1749

Erythrocytes
in amniotic fluid, technique for eliminating, 1568

Estriol
assays in pregnancies complicated by rhesus immunization, 1634
concentration in amniotic fluid, as index of fetal state, 1609
levels, and fetal well-being, 1566
values in mongolism, 2033

Estrogen
excretion in high-risk pregnancies, 1588
urinary excretion of in prolonged pregnancies, 1644

Family
attitudes toward MR, 2410
brothers and sisters of MR; psychological and sociological study, 2413
counseling parents of MR, 2414
developmental plans for MR in foster families, 2417
functioning, in cases of inst and non-inst MRs, 2405
guide for parents of MR, 2412
interactions with low adjusted, high adjusted, and MR members, 2419
MMPI scores of parents of MR, 2404
relations in cases of MR, 2298

Feeding Behavior
of MR child; role of nutritionist in, 2436
in MR child; role of occupational therapist in, 2437

Fertilization
and cleavage in vitro of preovulator human oocytes, 1632

Fetal Death
from intrauterine transfusion, in baboons, 1672

Fetus
abnormal placental development and health of, 1620
analyzing amniotic fluid to determine age of, 1649
antibodies produced in, from maternal antigen, 1696
B6 deficiency in rat; teratogenic effects, 1799

Fetus (continued)
cellular changes in, during growth, 1952
damage to, from environmental agents, 1638
determining sex of, by sex chromatin analysis, 1597
early developmental abnormalities in, 1932
effect of environment on, 1970
effect of folate acid deficiency on, 1714
effect of maternal phenylketonuria on rat, 1890
effect of trimethadione and paramethadione on, 2118
effect of tromethamine or bicarbonate infusions on circulation of, 1752
evaluation of, by plasma estriol measurements and ultrasonography, 1611
fetal distress and pH value of umbilical cord blood, 1915
feto-maternal transfusion as index of placental damage and fetal risk, 1605
immunologic mechanisms in, 1684, 1692
intrauterine visualization of, in hydramnios, 1573
low birth weight, 1957
monitoring of, during labor, 1640
monitoring techniques, 1600
small-for-date, hyperflexion of, 1967
small-for-date, radiological maturity of, 1966
state of, correlated with hyperbilirubinemia and meconium staining in mother, 1713
ultrasonics in intrauterine head measurement of, 1578
ultrasonics in monitoring development of, 1585
undernourished; technique for feeding orally, 1625

Fibroblasts
beta-glucosidase activity in, in Gaucher's disease, 1843
in cystic fibrosis, Hunter's syndrome and Hurler's syndrome, 1781
hexosaminidase deficiencies in, in ganglioside storage diseases, 1844

Folic Acid
effect of maternal deficiency of on fetus, 1714
serum-phenytoin levels in, 2030

France
PMR children institutionalized in, 2366
recommendations for dealing with multiple abnormalities in, 1559
rehabilitation of MR in, 2271

Galactosemia
carbohydrate excretion in, 1782
screening for, in New York State, 1802
unstable galactose-1-phosphate uridyl transferase, 1820

Gammaglobulin
effectiveness in rubella, 1677

Gangliosides
disorders of storage of, 1905
GM1 gangliosidosis, type 2, 1827
GM2 storage diseases; hexosaminidase in, 1844
juvenile GM2 gangliosidosis, 1790
metabolic disorders involving, 1814

Gaucher's Disease, 1907

Genetic Counseling: See Counseling

Genetics (See also **Hereditary Diseases**)
chromosome gain in higher plants, 2055
effect of chromosome redundancy on gene expression, 2054
influence of, on behavior, 2167
in Lowe's syndrome, 1835
mutagenicity of chemicals and drugs, 1639
prospects for genetic intervention in man, 1630
social impact of advances in, 1629
3 alpha genes, in apes, 1724

Globulin
Rh immunity globulin donor program in Massachusetts, 1631

Gluconeogenesis
hepatic, and lipogenesis in intrauterine growth retardation in rat, 1955

Glucosephosphatase Deficiency
von Gierke's disease, 1888

Glycine
excretion in nonketotic hyperglycinemia, 1871

Glycogen Storage Disease
surgical therapy for, 1887

Glycogenosis
detection of, in amniotic fluid, 1884

Great Britain (See also **England, Ireland**)
anencephalus in, 1971
Education Bill of 1971, 2205
residential occupational training facility for MR in Wales, 2278

Growth
abnormalities and maturation in MR, 2122
parameters of, in failure to thrive children, 1832
placental and fetal, cellular changes during, 1952
retardation in children, from hypoglycemia, 1860

Growth Disorders
characteristics of intrauterine growth retarded population, 1956
effect of emotional deprivation on, 2003
experimentally induced growth retardation in rat, 1955
fetal, and hyperflexion, 1967
fetal, and ossification delay, 1966
intrauterine growth retardation, 1969
testing for growth hormone abnormalities in, 1868
with thyroxine-binding globulin deficiency, 1894

Hair
abnormalities and inborn errors of metabolism, 1775

Hand
metacarpal sign as diagnostic index, 1595

Hartnup Disease
MR in, 1809

Health and Welfare Planning
multidisciplinary approach toward MR, 1563

Hearing Disorders (See also **Deafness**)
screening for neonatal, 1592

Heart Enlargement
and heart failure, associated with hypoglycemia, 1653

Hemangioma
diffuse neonatal hemangiomatosis, 1954

Hematologic Diseases
congenital hemolytic anemia, 1659

Hemoglobin
tablet for distinguishing between adult and fetal in vaginal bleeding, 1579

Hepatitis
following mononucleosis with Reye's syndrome, 1716

Hepatitis, Infectious
associated with mongolism, 2091, 2098
experimental exposure of MR to, 2376

Hepatolenticular Degeneration
Wilson's disease, observations and literature review, 1818
Wilson's disease, triethylene tetramine dihydrochloride in, 1906

Hereditary Diseases
arthrogryposis multiplex congenita, 1960
congenital gargoylism, 1829
congenital sensory neuropathy, 1953
cystinuria, 1817
epilepsy associated with hypoacusis and heterochromia iridis, 2010
little leopard syndrome, 1965
moyamoya disease, 1978
ophthalmoplegia and visuo-motor functions, 1972
sea-blue histiocyte syndrome, 1679
spastic paraparesis with sensory neuropathy, 1973

Herpesvirus Hominis
neurological manifestations and use of idoxuridine, 1683

Histidinemia
delayed histidine transamination, 1877
maternal, with normal birth, 1897
speech and language abnormalities in, 1902

Histiocytes
sea-blue, 1679, 1771

Homocystinuria
with neuroblastoma, excretion of cystathionine and homocystine in, 1840

Hormones
administration of testosterone and gonadotropin in Klinefelter's syndrome, 2044
growth hormone investigation in cases of MR and emotional disturbance, 1868
human chorionic somatomammotropin values in high-risk pregnancies, 1643
human growth, and pituitary myxedema, 1783
plasma growth, and chromosome abnormalities, 2047

Hungary
incidence of CNS malformations in, 1541

Hunter's Syndrome: See under Mucopolysaccharides

Hurler's Syndrome: See under Mucopolysaccharides

Hyaline Membrane Disease
adrenal cortex in, 1760
adrenal gland structure in, 1759
diagnosis and treatment, 1758
diagnosis of, by amniocentesis, 1591
lung phospholipids in, 1921
mechanical ventilation in, 1757
peritoneal dialysis in, in premature infants, 1939
therapy for, 1919, 1920

Hydramnios
with anencephalus, 1567, 1974
intrauterine fetal visualization in, 1573

Hydrocephalus
cerebrospinal fluid dynamics in, 1923, 1924
with dementia; ventriculovenous shunting in, 1958
from diffuse neonatal hemangiomatosis, 1954
EEG patterns of children with, 1935
metabolism of cerebrospinal fluid in, 1927
with stenosis of cerebral aqueduct, following mumps encephalitis, 1682
from toxoplasmosis, 1701
ventriculoatrial shunts in, 1936, 1958

Hyperbilirubinemia
Crigler-Najjar syndrome treated with phototherapy, 1736
in cytomegalic inclusion disease, 1657
in late pregnancy, and correlation with meconium staining, 1713
phenobarbitone in, 1702
phototherapy failure in, 1666
in premature infants, 1678
transient unconjugated, in monkeys, 1681

Hypercalcemia
idiopathic, associated with kidney hypoplasia, 1787

Hyperglycemia
associated with diarrhea, 1680

Hyperkinesis
intellectual ability and performance in, 2180

Hyperphenylalaninemia
mutant enzymes in, 1806
in phenylketonuria, 1804
urinary amino acid excretion in, 1819

Hypertension
renovascular, in children, 1913

Hypoglycemia
asymptomatic, in high risk newborn infants, 1735
in children, and growth retardation, 1860
in glycogen storage disease, 1887
of infancy, followed by diabetes mellitus, 1861
neonatal, diagnosis and etiology, 1734
neonatal, induced by drugs, 1652
neonatal, with cardiac enlargement and heart failure, 1652
with neonatal sepsis, 1668
in von Gierke's disease, 1888

Hypothyroidism
in children; prognosis, 1786
congenital and acquired, 1867
deiodinase deficiency in, 1857
effect on I.Q., school performance, and electroencephalogram pattern in children, 1900
pituitary, 1783
serotonin blood levels in, 1883

Hypoxia
tissue, and defective renal excretion, in infantile diarrhea, 1847

Immunity
autoimmune disorders and sex chromosome abnormalities, 2029

Immunoglobulins
in amniotic fluid, 1590
in detection of congenital infections and in immunization prophylaxis, 1613
G, in cerebrospinal fluid of children, 1614
IgA and IgE deficiency, with MR, 1669
levels of in amniotic fluid, and titers of specific antiviral antibodies, 1618
measles, in subacute sclerosing panencephalitis, 1698
response to immunofluorescence in rubella, 1674

Immunology
experimental antigen with maternal-fetal effect, 1696
for prevention of viral hepatitis, type B, 2376
in promotion of human fetus, 1692
unification of fetal and neonatal, 1684

Infant
at risk, registers for, 1544

Infant, Newborn
asymptomatic hypoglycemia in, 1735
behavioral assessment of, 2186
birthweight in hemolytic disease of, 1746
decreasing bilirubin levels in with orotic acid, 1695
diagnosis of histidinemia in, 1877
EEG of, and brain maturation, 1612
effect of defective birth on family, 2403
effect of sensory deprivation on, 2005
estimating gestational age of, 1603
hypoglycemia in, 1652, 1653, 1734
immunologic function in, 1684
intensive care center for, in Utah, 1636
low birth weight, effect of prenatal nutrition on, 1962
low birth weight, mortality in, 1647
low birth weight, prognostic study, 1940
multiple abnormalities in, and parental chromosome abnormalities, 2032
neurological criteria for estimating post-menstrual age of, 1610
prematurity trends in the United States, 1622
seizures in, 2009
sex ratio in toxemia of pregnancy, 1660
skull circumference as index of brain size in, 2123
social problems raised by pathological, 2420
test to distinguish differences among normal newborns, 2187
trisomy 18 in, 2056
very low birth weight, 1947, 1948
with X chromosome abnormalities; prognostic studies, 2082

Intelligence (See also **Cognition**)
academic achievement and minimal brain dysfunction in MR, 2183
in children, after acute brain injury, 1767
in children of interracial matings, 2139
in children with learning disabilities and controls, 2179
concept of, in special education, 2224
day care for culturally deprived children and development of, 2131
in deafness, testing for, 2190
among disadvantaged school children, 2129
distribution of, 1549
effect of home rearing on defective, 2418
effect of hypothyroidism on, in children, 1900
effect of malnutrition on, 1855, 1909
effect of poverty on, 2321
effect of pre- and postnatal environment on, 2145
effect of socioeconomic factors on, 2143
factors adversely affecting development of, 1562
genetic and environmental factors in IQ, 2146, 2147, 2148, 2149, 2150
IQ changes in hospitalized MR, 2153
kinesio-perceptual test battery correlated with, 2128
and learning in epilepsy, 2025
loss, in phenylketonuria, 1794
and perceptual-motor performance in hyperkinetic child, 2180
socioeconomic factors in development of, 2141
study of relative heritability of, 2127

Intelligence Tests
administered to Puerto Rican working class children, 2181
Bayley Infant Scales of Mental and Motor Development, 2138
Denver Developmental Screening Test, 2129
role of language training in improving scores on, 2208

Iran
study of anencephaly in, 1949

Ireland
treatment of MR in, 2387

Israel
anencephalus and spina bifida in, 1942

Japan
study of sex chromosome abnormalities in, 2062

Jaundice, Neonatal (See also **Hyperbilirubinemia**)
bilirubin levels in, 1691
phenobarbital in, 1667
phototherapy in, 1726
related to breast milk, 1697

Keloid
rare formation, in Rubinstein-Taybi syndrome, 1951

Kernicterus
relationship between free bilirubin level and, 1693

Klinefelter's Syndrome
administration of testosterone and gonadotropin in, 2044

Krabbe Leukodystrophy: See **Cerebral Sclerosis, Diffuse**

Kuf's Disease: See under **Amaurotic Familial Idiocy**

Kwashiorkor
followup study of South African children with, 1845

Labor
difficulty in and resultant neurological conditions, 1765
monitoring of fetus during, 1584, 1640

Language
abilities in disadvantaged children, 2201
behavior measured in normal and EMR children, 2178
development in SMR and normals, 2168
as maturational process, 2171
in mildly MR; sociocultural aspects, 2163
role of nonprofessional in teaching language to MR, 2208
sign language for nonverbal child, 2245
and speech abnormalities in histidinemia, 1902

Laterality
effects of age and mongolism on handedness, 2115

Laurence-Moon-Biedl Syndrome
and Lesch-Nyhan syndrome, in same family, 1934

Lead
method for detecting in paint, 1715

Lead Poisoning
in children, in New York City, 1688
in children, screening program for, 1706
diagnosis and therapy, 1727
public health measures against, 1661
test for use in screening campaigns to identify, 1715

Learning
assessing capacity for in MR, and development through education, 2200
and attention, in MR, 2134
behavior therapy with nonverbal psychotic child, 2251
deficits and activity changes in postencephalitic rats, 1707
discrimination, of MR, 2156
effect of learning method on learning stage in MR and normal adolescents, 2204
encoding processes and implicit verbal associations in MR, 2152
facilitated by meaningful material, 2202
Federal funds for disability programs, 2320
intelligence in children with disabilities in, 2179
and intelligence in epilepsy, 2025
matching to sample behavior, with touch tutor, 2228
mechanisms in color sorting discrimination, in normals and SMR, 2155
problems, course to help teachers identify children with, 2235
television as stimulus for acquiring reading skills, 2234
transfer of stimulus control in SMR, 2154
verbal and motor tasks, in TMR, 2231

Learning Disorders
in disadvantaged children, 2182
neurological learning disability syndrome, 2114
related to hypothyroidism, in children, 1900

Learning Disorders (continued)
the slow learner, 2195, 2196, 2197
utility of medical evaluation in, 1762

Legal Status of MR
criminal responsibility, 2338
in England, 2313
financial support available to MR, in Belgium, 2325
human rights, 2327

Legislation
Developmental Disabilities Services and Facilities Construction Act, 2310, 2312, 2330
Developmental Disabilities Services and Facilities Construction Amendents of 1970, 2331
Federal funds for learning disabilities programs, 2320

Lesch-Nyhan Syndrome
altered kinetic properties of mutant enzyme in, 1893
effect of allopurinol on pyrimidine metabolism in, 1885
enzyme activity in, 1780
hypoxanthine-guanine phosphoribosyltransferase deficiency in, 1777, 1800
and Laurence-Biedl syndrome, in same family, 1934
prenatal diagnosis and screening for, 1801
preventive control of, 1816
testing for hyperuricemia, 1580
using hair follicles for detection of heterozygotes in, 1838

Limbs
abnormalities of, and MR, 2113
multiple congenital anomalies of, with microcephaly, 1943

Lipid Metabolism, Inborn Errors
cerebral lipidosis, 1796
infantile neurolipidosis and failure of myelination, 1795
Wolman's disease with hypolipoproteinemia, 1792
1-cell diseases, 1826

Lipochondrodystrophy
congenital gargoylism, 1829
gargoylism; 3 cases, 1852

Lipoma
associated with MR, 1984

Lithium
in pregnancy, recommendations, 1690
and safety of fetus during pregnancy, 1703

Lowe's Syndrome: See under **Abnormalities, Multiple**

Lysergic Acid Diethylamide
effect on chromosomes, 2079

Lysosomes
in pathology of nervous tissue, 1810
types of congenital abnormality in, 1813

Malnutrition
associated with social class, 2145
behavioral measurement of, 1858
and brain development, 1898
and brain development, in guinea pig, 1851

Malnutrition (continued)
effect on animal growth, 1899
effect on intellectual functioning, 1909
effect on physical and mental development, 1845
fetal, and maternal leukocyte metabolism, 1846
fetal, functional effects, 1805
fetal, oral feeding of fetus in, 1625
and growth retardation, 1969
physical and mental effects of, 1854, 1855
prenatal, and low birth weight infant, 1962

Maple Syrup Urine Disease
dietary intervention in, 1910
thiamine treatment for, 1850

Marriage
and MR, 2169

Massachusetts
Rh immune globulin donor program in, 1631

Measles
mass immunization for, 1733

Memory
recall and material associative strength in MR, 2135

Meningitis, Viral
prognosis in, 1748

Metabolism (See also **Metabolism, Inborn Errors; Carbohydrate Metabolism, Inborn Errors; Lipid Metabolism, Inborn Errors**)
bilirubin, proteins Y and Z in, 1689
calcium-phosphate, in Turner's syndrome, 2074
carbohydrate, in experimental intrauterine growth retardation in rats, 1889
of cerebrospinal fluid, in hydrocephalus, 1927
of dermatan sulphate or heparan sulphate, in mucopolysaccharidoses, 1811
of drugs by feto-maternal system, 1723
in mammalian synaptosomes, 1626
of tyrosine, in phenylketonuria, 1803

Metabolism, Inborn Errors (See also **Carbohydrate Metabolism, Inborn Errors; Lipid Metabolism, Inborn Errors**)
accumulations and deficiencies of gangliosides, 1814
age related analysis of glucose-6-phosphate deficiency, 1821
Alexander's disease, 1770
aspartylglycosaminuria, 1812
congenital lactic acidosis in two siblings, 1834
Fabry's disease, 1788
gamma-glutamyl transpeptidase deficiency with MR, 1822
and hair abnormalities, 1775
hydroxylysinuria, 2051
hyperphosphatasia, with cataract and MR, 1773
hyperprolinemia, 1784, 1839
lipofuscin storage disease, 1977
myelin sheath defect in leukodystrophy, 1976
nonketotic hyperglycinemia, 1871
pathology of hepatobiliary diseases, 1785
prenatal detection of, 1870
pycnodysostosis, 1791
renal reabsorption of monoamino monocarboxylic acids, 1809
renal tubular reabsorption of amino acids, 1808

Metabolism, Inborn Errors (continued)
sarcosinemia, with MR, 1793
test for glycogen storage diseases, 1593
of tryptophan, in mongolism, 2052
vitamin metabolism in childhood disorders, 1577
Wilson's disease, 1818

Michigan
university training for nutrition personnel in, 2435

Microcephaly
with cytomegalic inclusion disease, 1747
and MR, 2119
with multiple congenital anomalies of limbs, 1943

Microphthalmos
sex-linked, with deafness and MR, 1925

Minimal Brain Dysfunction: See **Brain Damage, Chronic**

Minnesota
special education for MR in, 2342

Molecular Biology
cellular membranes in MR, 1626
cellular organelles and membranes in MR, 1552

Mongolism
amylase heterogeneity in, 2061
associated with infectious hepatitis, 2098
behavior therapy for mongoloid boy, 2255
biochemical abnormalities in, 2052
congenital, 2031
congenital heart disease in, 2060
dermatoglyphics as index of, 2040
diagnosis of, with dermatoglyphic nomogram, 2108
due to maternal mosaicism, 2090
effect of, on laterality, 2115
familial predisposition to thyroid disease in, 2081
fetoplacental function in, 2033
foster family care in, 2411
incidence of Australia antigen in, 2083
and infectious hepatitis, 2091
language development in, 2168
language learning in, 2171
mosaic parents of mongol propositi, 2065
with nonketotic hyperosmolar diabetic coma, 2043
orienting reflex study in, 2126
with phenylketonuria, 1824
prenatal diagnosis of, by X-ray, 2100
response to atropine in, 2103
RNA synthesis by leukocytes in, 2053
study of mental and motor development in, 2138
with XYY:48,XYY,G+, 2087

Mosaicism
in Lesch-Nyhan syndrome, 1816
maternal, in mongoloid births, 2090
monosomy G with, 2073
mosaic parents of mongol propositi, 2065
tetraploid, and therapeutic abortion, 1582
45,XX,21-46,XX,21, with monosomy 21, 2072

Mother-Child Relations
infantilization of MR, 2409
psychotherapy for mothers of MR, 2407

Motivation
effect on reminiscence in MR, 2133
of parents in repeat evaluation of MR children, 1586

Motor Skills
deafness, and psychic development, 2117
deficiency of, in congenital rubella, 1725
kinesio-perceptual testing, 2176
perceptual-motor survey of MR, 2130

Movement Disorders
naturally occurring, need for reevaluation, 2111

Mucopolysaccharides
biochemistry of mucopolysaccharidoses, 1811
in cystic fibrosis, Hunter's syndrome and Hurler's syndrome, 1781
detection of, in urine, 1879
Hunter's syndrome, in British Columbia, 1852, 1862
Hurler's syndrome, 1807, 1825, 1830, 1874
Hurler's syndrome, in British Columbia, 1862
metachromatic leukodystrophy with mucopolysaccharidosis, 1853
molecular weights of, in brain and other organs of patients, 1874
mucopolysaccharidosis, type 1, 1831
a new mucopolysaccharidosis, 1863, 1864, 1865
rare form of mucopolysaccharidosis in child, 1872
relative frequency of Hunter and Hurler syndromes, 1880, 1881, 1882
Sanfilippo syndrome, 1825, 1828

Muscles
absent abdominal, with CNS involvement, 1968

Nerve Tissue
lysosomes in pathology of, 1810

Netherlands
sheltered workshop in, 2281

Neuroblastoma
familial, and trisomy 13, 2086

Neurofibromatosis
hemihypertrophy in, 1911
in renovascular hypertension, 1913
von Recklinghausen's, 1912

Neuroglia
astroglial enzyme defect, with MR, 1770

Neurologic Manifestations
with acrocentric supernumerary chromosome, 2048
associated with sea-blue histiocytes, 1771
and biological abnormalities, in autism, 1997
in Cockayne's syndrome, 1917
correlated with behavior, 2166
in diffuse neonatal hemangiomatosis, 1954
in hepatobiliary diseases, 1785
infantile neurolipidosis and failure of myelination, 1795
in mice, from glutamate, aspartate, and cysteine, 2112
neurocutaneous syndrome, seizures and MR, 1946
neurological learning disability syndrome, 2114
orthomolecular therapy in, 2309
from radiation, physical factors, viruses, and drugs, 2064
in reading disabilities, 2229
resulting from obstetrical complications, 1765
spastic paraparesis with sensory neuropathy, 1973
tests for diagnosing and treating, 1617
viral infections in, 1983

Neurologic Manifestations (continued)
visual evoked responses in MR and normal subjects, 2136

Neurons
accumulation of smooth cisterns, multivesicular bodies and zebra bodies in, 1876

Neuroses
in MR, 2159

Nevus
sebaceous of Jadassohn, with MR, 1946

New Guinea
iodine injections to prevent cretinism in, 1849

New York
lead poisoning among children in New York City, 1688
Project UNIQUE for EMR (Rochester), 2226
screening for galactosemia and phenylketonuria in, 1802

New Zealand
Intellectually Handicapped Children's Society, 2263, 2317
opportunity workshop in, 2266

Niemann-Pick Disease, 1907

Nursing
effect of, on family of defective newborn, 2403
home care for MR, 2301
in interdisciplinary nutritional program, 2439

Nutrition
manpower needs in field of, 2434
nutritionists in University Affiliated Centers, 2440
prenatal, 1957
of purine biosynthesis, 1772
role of, in prevention and management of MR, 1561
university training for personnel, 2435

Operant Conditioning: See **Behavior Therapy**

Ovary
ovarian dysgenesis, in trisomy 18, 2075

Parent-Child Relations
group therapy for parents of handicapped children, 2408
home background of SMR compared to normal children, 2415
in MR, 2406

Parents
necessity for involvement of in MR inst, 2383

Paresis
spastic, with sensory neuropathy, 1973

Perception
deficits, in congenital rubella, 1725
Frostig program for visual perception development, 2223
kinesio-perceptual testing, 2176
object concept in deviant children, 2142
perceptual-motor survey of MR, 2130
in symptomatic autism, 2000
visual presentation and recognition, 2132
and visuo-motor functions in congenital ophthalmoplegia, 1972

Personality Assessment
of inst MR, 2172

Phenylalanine
as inhibitor of uptake of amino acids, 1903
turnover levels in mild and severe PKU, 1833

Phenylketonuria
amino acid diet for, 1776
in children, cognitive development in, 2144
diagnosis and prognosis in, 1901
diagnosis and treatment, 1878
dietary intervention in, 1910
effect of low phenylalanine diet on, 1886
effect of maternal, on rat fetus, 1890
false negative screening tests, 1895
intelligence loss in, 1794
maternal, during pregnancy, 1842
mild and severe; comparative studies in 2 infants, 1833
with mongolism, 1824
mutant enzymes in, 1806
screening for, during pregnancy, 1841
screening for, in New York State, 1802
sex ratio in occurrence of, 1778, 1779
tyrosine metabolism in, 1803
urinary amino acid excretion in, 1819
in 10 Egyptian families, 1804

Phospholipids
in blood cells, comparative study, 1670
in hyaline membrane disease, 1591, 1921

Phototherapy
failure in hyperbilirubinemia, 1666
in hereditary hyperbilirubinemia, 1736
in neonatal jaundice, 1726
and skin color, 1663, 1664

Physical Education and Training
value for MR, 2207

Physical Therapy
role of, in treating MR, 2438

Pigmentation Disorders
incontinentia pigmenti achromians, 1933

Placenta
cellular changes in during growth, 1952
localization of, by isotopic placentography, 1598
methods of localization of, 1572
pathology of, in hydrops fetalis, 1721
transfer of diphenylhydantoin by, 1704

Poland
residential facilities for MR in, 2323

Polyploidy
in amniotic fluid cells, 1575

Potassium
deficiency in epilepsy, 2017

Prader-Labhardt-Willi Syndrome: See under **Abnormalities, Multiple**

Pregnancy
action line method for management of rhesus isoimmunization in, 1731
aminoglutethimide in, 1717
cytohormonal and interhormonal relationships in, 1564
estriol surveys in rhesus negative women, 1634
high-risk, estriol levels in, 1566
high-risk, estrogen excretion in, 1588
high-risk, human chorionic somatomammatropin levels in, 1643
high-risk, medical services in, 1619
inadvertent rubella virus vaccination during, 1753
lithium therapy during, 1690, 1703
optimum time for amniocentesis during, 1583
phenylketonuria during, 1842
polyhydramnios in, 1963
prediabetic, free fatty acid level in, 1642
protein concentrations in Rh negative blood in, 1687
in pseudopseudohypoparathyroidism, 1892
Rh sensitized, amniotic fluid spectrometry in management of, 1615
rubella preceding, as possible cause of congenital malformations, 1737
screening for phenylketonuria during, 1841
testing for rubella during, 1729, 1730
thyroid replacement therapy in and effect upon children, 1866
twin conceptions and fetal disorders, 1620
uterine bleeding in, and congenital defects, 1623

Proline
in hyperprolinemia, 1839
low proline diet in type 1 hyperprolinemia, 1896

Proteins
in fetal erythroblastosis, 1738
hepatic Y and Z, in bilirubin metabolism, 1689
role of, in cerebral adaptation, 1627
synthesis of, role of lysosomes in, 1813
Y and Z, in neonatal jaundice, 1681

Pseudopseudohypoparathyroidism
pregnancy in, 1892

Psychiatry (See also **Psychotherapy**)
child, role in MR, 2423
child, socioeconomic background of therapist in, 2424
in diagnosis and treatment of MR, 1599
for MR adults, by television, 2296

Psychological Tests
distribution of reaction times in MR, 2140

Psychosis (See also **Autism**; **Schizophrenia, Childhood**)
in MR, 2159

Psychotherapy (See also **Psychiatry**)
in childhood schizophrenia; use of video tape to document behavioral change, 2300

Psychotherapy, Group
for mothers of MR, 2407
for parents of handicapped children, 2408
for self-induced seizures in epilepsy, 2299

Purines
metabolism of, and MR, 1772

Pyridoxine
deficiency, diagnosed by tryptophan-tolerance test, 2026

Pyrimidines
metabolism of, in Lesch-Nyhan syndrome, 1885

Radioautography
in diagnosis of chromosome anomalies, 2073

Recklinghausen's Disease of Nerve: See under **Neurofibromatosis**

Recreation, 2396
camping for MR, 2395
holiday camp for MR, 2399
horseback riding for MR, 2288, 2400
library services for MR, 2357, 2358, 2389
mountain outing for handicapped, 2401
municipal programs for MR, 2398
public library services for MR, 2359, 2360
therapeutic, for disabled children, 2397
toy library for MR, 2393

Refsum's Syndrome
diagnosis, etiology, and treatment, 1891

Rehabilitation, 2343
drama as therapy for MR, 2284
in England, recommendations for, 2318
evaluation of care and services, 2344
foundations for a science of, 1545
interdisciplinary clinical services, 2334
normalization principle in, 2238, 2363, 2364
preparing long-term MR residents for community placement, 2346
as priority in MR, 1551
teamwork for MR, 2260
training of therapists engaged in child health and, 2432
volunteer and parent participation in MR institutions, 2367

Rehabilitation, Vocational, 2267
for brain injured adolescents; abuses, 2349
counseling in sheltered workshops, 2272
design and testing of sheltered workshop, 2250
economic benefits of, 2268, 2332
Flame of Hope sheltered workshop program, 2275
functions of sheltered workshop supervisors, 2273
hiring MR from sheltered workshops, 2261
hospital work-study program for EMRs, 2280
for inner city MR, 2340
for MR, in France, 2271
for MRs who repeatedly failed in labor market, 2276
for multiply handicapped blind students, 2277
occupational therapy assistants, 2422
opportunity workshops, 2263
remedial education to prepare for vocational goals, 2282

Rehabilitation, Vocational (continued)
sheltered employment, 2265
sheltered workshop for SMR, 2269
sheltered workshop in Australia, 2259
sheltered workshop in the Netherlands, 2281
sheltered workshops for MR, 2270
Taranaki (New Zealand) opportunity workshop, 2266
teaching child care to handicapped children, 2262
Vocational & Rehabilitation Research Institute (Calgary, Canada), 2274
vocational adjustment rating scale for MR, 2283

Religion
for educable and trainable MR, 2336
role of chaplains in MR institutions, 2371

Research
multidisciplinary efforts in MR, 1548
priorities for MR, 1546, 1547, 1549, 1550, 1551, 2315, 2423

Residential Facilities
admission of autistic children to MR hospitals, 2377
bedroom groupings of handicapped, in England, 2368
Bevan Lodge, Courtenay, B. C., Canada, 2382
in England, 2379
in England, assessment of, 2362
in England, mistreatment of MR in, 2381
in England, recommendations for, 2318
evaluation of demand for short- or long-term care of MR (England), 2373
factor analysis of MR admissions to, 2369
factors influencing decision to commit MR, 2405
hostels for MR, 2375
IQ changes in MR in, 2153
Kinsmen Elmwood Lodge, Saskatoon, Saskatchewan, Canada, 2380
length of MR stay in, in England, 2378
library services for MR in, 2357, 2358, 2389
mortality of MR in (Sweden), 2374
for MR, in Denmark, 2361
for MR, in Ireland, 2387
for MR, in Poland, 2323
necessity for parental involvement in, 2383
normalization principle in, 2238, 2363, 2364
peer groups among inst MR, 2370
PMR children in, in France, 2366
program planning for deaf MR children in, 2392
providing occupational training for MR, in Wales, 2278
rating behavioral characteristics of MR in, 2172
recommended standards for library services in, 2390
resident intervention groups in, 2391
role of attendant level personnel in, 2386
role of chaplains in, 2371
role of parent and volunteer workers in, 2367
Salmon unit for MR, 2384
short-term care hospital for MR, 2388
the state mental institution of the future, 2385
versus family care, in mongolism, 2138

Respiratory Distress Syndrome
assessing amniotic fluid to prevent, 1608
mechanical ventilation in, 1755

Retinitis Pigmentosa
with MR, deafness, and XX/XO sex chromosomes, 2067

Rh Factors
birthweight in hemolytic disease of newborn, 1746
hemostatic failure associated with rhesus isoimmunization, 1740
Rh immunization in spontaneous abortion, 1720
role of phenobarbitone in isoimmunization, 1650

RNA
synthesis by leukocytes in mongolism, 2053

Rubella
antecedent to gestation period, as possible cause of congenital malformations, 1737
antibody determination among pregnant women, 1743
antibody levels in gamma globulin preparations, 1856
associated with chromosome abnormalities, 2095
congenital, incidence of in Belgium, 1718
congenital, mental development of children with, 1739
congenital, motor deficits in, 1725
and diastrophic dwarfism, 1665
distinguished from rubeola, 1732, 1733
effectiveness of gammaglobulin in, 1677
immunization program, 1728
immunoglobulin responses in, 1674
inadvertent inoculation of pregnant women for, 1710, 1753
laboratory diagnosis of, 1676
mass vaccination for, in England, 1751
noncontagiousness of Cendehill vaccine strain for, 1711
policy for vaccinating children for, 1671
postpartum vaccination for, 1685, 1742
prevention of, 1675
retrospective survey of, in Washington State, 1705
subclinical, testing pregnant women for, 1729, 1730
vaccination for, 1651
vaccination for, during puerperium, 1741
vaccination for, results of program, 1708
vaccines, exposure of susceptible pregnant women to, 1709
virus vaccines studied during puerperium, 1744

Rubenstein-Taybi Syndrome: See under **Abnormalities, Multiple**

Sanfilippo's Syndrome: See under **Mucopolysaccharides**

Scandinavia
genetic counseling in, 1596

Schizophrenia, Childhood (See also **Autism**)
behavioral changes documented by video tape in, 2300
serotonin uptake by platelets in, 2001

Self Concept
of MRs, compared with normals, 2162

Self-Mutilation
in de Lange syndrome, 1931

Sensory Deprivation
effect of, on newborn infant, 2005

Serotonin
blood levels in hypothyroidism, 1883
uptake in blood platelets, in childhood schizophrenia, 2001

Sex Chromosome Abnormalities
and autoimmune disorders, 2029
excess Barr bodies in Swedish MR population, 2063
Japanese study of, 2062
Renpenning syndrome, 2034
XYY and crime, 2030
Y heterochromatin and XX males, 2050

Skin
delayed type reactions of in ataxia telangiectasia, 1986
extra posterior cervical, as indicator of chromosome abnormalities, 2035
galvanic skin response in MR, 2121

Slovenia
study of retarded delinquents in, 2165

Social Adjustment
of MR, 2164, 2246
peer groups among inst MR, 2370
sex and the MR, 2240, 2241

Socioeconomic Factors
adverse, and competence formation, 2002
assessment of, in predicting low birth weight infants, 1647
the deprivation syndrome, 1999
in development of intelligence, 1854, 2131, 2141
effect on intelligence, 2143
in EMR, 2185
in epidemiology of MR, 1553
ethnic distribution of children in EMR and LD classes, 2211
in etiology of MR, 1645
in intelligence, and testing of socioculturally deprived children, 2184
in language of mildly MR, 2163
poverty and intelligence, 2321
psychosocial deprivation and infant development, 2160

Spectophotometry
amniotic fluid, in anencephalic pregnancies, 1567
amniotic fluid, in management of Rh sensitized pregnancy, 1615

Speech
development and voluntary breath control in MR, 2124
relevance of feeding sequence to development of, 2308
therapy for disorders, 2293

Sphingolipids
chemical techniques in diagnosing sphingolipidoses, 1815

Spina Bifida, 1918, 1928
coordinated care for, 1959, 2290, 2335, 2341
epidemiology of, 1926, 1941
in Israel, 1942
with urinary ascites, 1964

Statistics
correlation of perinatal factors and MR, 2116
occurrence of minor congenital malformations unassociated with major congenital malformations, 1633

Steroids
pregnene-3(x),20(B)-diol in neonatal jaundice, 1697
in treatment of hyaline membrane disease, 1759, 1760

Subacute Sclerosing Panencephalitis
measles immunoglobulins in, 1698

Suicide
attempts among MR in institutional population, 2158

Sulfatases
A, in metachromatic leukodystrophy, 1990

Sweden
mortality of inst MRs in, 2374
normalization for MR in, 2316, 2324
study of excess Barr bodies in MR population, 2063

Switzerland
study of congenital malformations in, 1555

Synaptic Vesicles
membrane potentials and metabolic performance in mammalian synaptosomes, 1626

Tay-Sachs Disease: See **Amaurotic Familial Idiocy**

Teaching
art education of MR, 2192
certification for teachers of MR, 2429
intelligence and personality testing in, 2215
machine, response of SMR to, 2194
MR reactions to school curriculum and implications, 2230
self-feeding skills to PMR by behavior therapy, 2252
under reorganization of MR care in England, 2314

Telangiectasia, Hereditary Hemorrhagic
moyamoya disease, 1978

Testing
ability to reason in EMR, 2185
attitudes in MR, by semantic differential, 2242
behavioral assessment in infants, 2186
Bender-Gestalt performance of organic and functional MRs, 2173
Cambridge Newborn Behavioral and Neurological Scales, 2187
correlation between frustration and bodyrocking in SMR, 2157
in diagnosis and treatment of neurological disorders, 1617
First Grade Screening Test, 2189
individualized performance profiles for MR, 2214
intelligence and personality, value of, 2215
kinesio-perceptual, 2128, 2176
neurological criteria for estimating post-menstrual age of newborn, 1610
orienting reflex response in mongolism, 2126
perceptual-motor survey of MR, 2130
in predicting educability of cerebral palsied children, 2177
psychomotor, of socioculturally deprived children, 2184
psychomotor retardation, by Brunet-Lezine development test, 2191

Testing (continued)
reminiscence in MRs and normals; manual crank test, 2133
urine, for mucopolysaccharides, 1879
vigilance and verbal conditioning in MR, 2134
Wechsler-Bellevue Full Scale Test, 2174

Thalassemia
3 alpha genes, in apes, 1724

Therapeutics (See also Drug Therapy)
for acidosis, 1750
amino acid diet for phenylketonuria, 1776
combined clinics for spina bifida, 2335
comprehensive care program for MR, 2322
coordinated care for spina bifida patients, 2290, 2341
diagnosis and treatment of MR, 1599
effect of on crime patterns of MR, 2161
emerging conceptions of mental illness and, 2333
enzyme replacement in metachromatic fibroblastic leukodystrophy, 1988
for epileptic MR, 2011
folic acid treatment in epilepsy, 2021
Foster Grandparent Program for MR, 2337
for glycogen storage disease, 1887
for herpesvirus hominis encephalitis, 1683
in hyaline membrane disease, 1758, 1919, 1920
in hydrocephalus with dementia, 1958
for hypoglycemia, 1734
in hypothyroidism, ameliorative and preventive, 1786
manual sign language programming for deaf MR, 2203
mattress for handicapped child, 2287
mechanical ventilation in hyaline membrane disease, 1757
methyldopa in Bartter's syndrome, 1789
orthomolecular therapy for brain injured or neurologically handicapped, 2309
parent-staff communication in treatment and management of cerebral palsy, 2307
peritoneal dialysis in hyaline membrane disease, 1939
phenobarbital in neonatal jaundice, 1667
phenobarbitone in rhesus hemolytic disease, 1650, 1658
in phenylketonuria, 1886
for polyhydramnios, 1963
recreation for disabled children, 2397
reinforcement therapy for SMR, 2289
in respiratory distress syndrome, 1755
sectioning of corpus callosum in, 2014
for speech disorders, 2293
triethylene tetramine dihydrochloride in, 1906
in type 1 hyperprolinemia, 1896
use of N-benzyl-beta-chloropropionamide in child psychiatric clinic, 2285
ventriculoatrial shunts in hydrocephalus, 1936
for von Gierke's disease, 1888
walker for child with ataxia, 2297

Thiamine
in treatment of maple syrup urine disease, 1850

Thumb
in trisomy 18, 2084

Thyroid
effect of maternal replacement therapy upon children, 1866

Thyroid Diseases
familial, in mongolism, 2081
twinning, and chromosome abnormalities, 1798

Toxemia
of pregnancy, and sex ratio of newborn, 1660

Toxoplasmosis
congenital, late onset of disease in, 1701

Trisomy, 2107
C(47XXC+), 2071
D (47,XXD+), with cyclops, 2101
partial C (6-12), 2109
X, with pycnodysostosis, 1791
13, and familial neuroblastoma, 2086
18, in Australia, 2028
18, incidence in MR research unit, 2069
18, incidence in newborn, 2056
18, incidence of, 2088
18, the thumb in, 2084
18, with ovarian dysgenesis, 2075
21, in mother and child, 2031
21, with hydroxylysinuria, 2051
21, with XYY, 2087
6-12 (47,XX,C+), 2073

Tuberous Sclerosis
and MR without adenoma sebaceum, 1930

Turner's Syndrome
calcium-phosphate metabolism in, 2074
clinical and chromosomal patterns in, 2027
diagnosed later as Cornelia de Lange syndrome, 1961
with XY chromosome constitution, 2094

Ultrasonics
in fetal cephalometry, 1578
safety of, in monitoring fetal development, 1585

Umbilical Cord
acid-base balance in fetuses with cord around the neck, 1914
blood, and fetal distress, 1915

United Kingdom
amniocentesis in, 1569

Uric Acid
levels in mongolism, 2052
in screening for Lesch-Nyhan and other syndromes, 1580

Utah
intensive care center for neonates, 1636

Vaccination
of children, for rubella, 1671
noncontagiousness of Cendehill vaccine strain of rubella virus, 1711
postpartum immunization to rubella, 1742
for rubella, 1651
for rubella, during puerperium, 1741
for rubella, exposure of susceptible pregnant women to vaccines, 1709
for rubella, in England, 1751

Vaccination (continued)
for rubella, inadvertent inoculation of pregnant women, 1710
for rubella, results of program, 1708

Virginia
incidence of anencephalus in central, 1929

Viruses (See also **Cytomegaloviruses; Encephalitis Viruses**)
associated with febrile convulsions, 1655
in chronic and degenerative diseases, 1983
encephalitis, effect on learning in rats, 1707
Epstein-Barr, in case of mononucleosis with Reye's syndrome, 1716
measles, injected in hamsters and followed by encephalitis, 1686
rubella vaccine, studied during puerperium, 1744

Vitamins
B6, and excretion of cystathionine and homocystine, 1840

Vitamins (continued)
B6 deficiency; teratogenic effects in rats, 1799
metabolism of, in childhood diseases, 1577

Voluntary Services
for MR, in Canada, 2353

Voluntary Workers
citizen advocates for MR, 2355
training paraprofessionals for work with MR, 2425

Von Gierke's disease: See **Glucosephosphatase Deficiency**

Washington
epidemiology of MR in, 1553
population characteristics of MR schools in, 2372
retrospective survey of rubella in, 1705

Wilson's Disease: See **Hepatolenticular Degeneration**

☆ U.S. GOVERNMENT PRINTING OFFICE : 1973—O-489-526

Mental Retardation Abstracts

Questions about the Division of Developmental Disabilities (except submission of information on current projects) should be addressed to:

Division of Developmental Disabilities
Rehabilitation Services Administration
330 C Street, S. W.
Washington, D. C. 20201

Change of address or removal of name

Please affix Abstracts mailing label here and mail to Mental Retardation Abstracts, Superintendent of Documents, Subscription Office, Government Printing Office, Washington, D.C. 20402

Fill in below only for change of address.
Please print clearly.

Name First Initial Last

Organization (leave blank if none)

Number and Street Address

City State

Country (if other than U.S.) Zip

I wish to discontinue receiving this publication.
Remove my name from the list.

DEPARTMENT OF
HEALTH, EDUCATION, AND WELFARE
SOCIAL AND REHABILITATION SERVICE
REHABILITATION SERVICES ADMINISTRATION
DIVISION OF DEVELOPMENTAL DISABILITIES
330 INDEPENDENCE AVENUE SW.
WASHINGTON, D.C. 20201

OFFICIAL BUSINESS

POSTAGE AND FEES PAID
U.S. DEPARTMENT OF HEW
HEW-392

U.S. DEPARTMENT OF HEALTH, EDUCATION, AND WELFARE

Social and Rehabilitation Service

Rehabilitation Services Administration • Division of Developmental Disabilities